

T H E S I S

on

A CLINICAL STUDY of the IMMEDIATE MANIFESTATIONS and
the LATER EFFECTS of ACUTE JUVENILE RHEUMATISM in
EDINBURGH and SOUTH EAST SCOTLAND.

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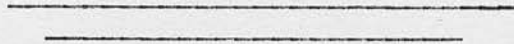


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A Clinical Study of the Immediate Manifestations and
the Later Effects of Acute Juvenile Rheumatism in
Edinburgh and South East Scotland.

INTRODUCTION.

The Frenchman, Guillaume de Baillou, at the beginning of the 17th century, is credited with being the first author to employ the term "rheumatism" as a substitute for "arthritis", a term which dates back to Hippocratic times. It was not until the year 1685, however, that Sydenham described in vivid language the disease which we recognise today as acute rheumatism, or rheumatic fever. The clinical picture painted by Sydenham is a model of descriptive genius wherein accuracy of observation has as its medium a most attractive and lucid exposition.

"This disease happens at any time, but especially in the Autumn, and chiefly affects such as are in the prime of life it begins with a chillness and shivering which are soon succeeded by heat, restlessness, thirst, and the other concomitants /

concomitants of a fever; in a day or two, and sometimes sooner, there arises an acute pain in some or other of the limbs, especially in the wrists, shoulders and knees; which shifting between whiles, affects these parts alternately, leaving a redness and swelling in the part last affected."

A year later, in 1686, Sydenham described chorea, although apparently he did not associate this disease with rheumatism. The clinical picture which he drew of the choreic child is remarkably graphic. "This disorder is a kind of convulsion which seizes children of both sexes from the tenth to the fourteenth year; it manifests itself by a halting or unsteadiness of one of the legs, which the patient draws after him like an ideot. If the hand of the same side be applied to the breast, or any other part of the body, the child cannot keep it a moment in the same posture, but it will be drawn into a different one by a convulsion, notwithstanding all his efforts to the contrary. Before a child who hath this disorder can get a glass or cup to his mouth, he useth abundance of odd gestures; for he does not bring it /

it in a strait line thereto, but his hand being drawn sideways by the spasm, he moves it backwards and forwards till, at length the glass accidentally coming nearer his lips, he throws the liquor hastily into his mouth, and swallows it greedily as if he meant to divert the spectators."

It was actually not until a century later, in 1788, that David Pitcairn first drew attention to the danger of cardiac complications occurring in the course of rheumatic fever.

Until the end of the eighteenth century rheumatism was regarded as essentially a disease of the articular structures, but early in the nineteenth century Balfour and Charles Scudamore took a wider view of the subject which formed the basis of our modern conception of the disease. These observers regarded rheumatism as being a disease of the cellular membrane, more particularly parts of the body having a fibrous texture, and it was along the lines of this hypothesis that they explained inflammation of the pericardium occurring in the course of the disease. The view of Scudamore is clearly revealed when /

when he writes: "There is not, probably, a more dangerous form of disease than a sudden seizure of the heart during the inflammatory state of the system in acute rheumatism."

In 1831 Bright drew attention to the relationship between chorea and acute rheumatism and observed the frequency of cardiac murmurs in chorea. In 1836, Bouillaud published his famous 'Law of Coincidence' between the occurrence of heart disease and acute rheumatism.

Sir Thomas Watson, in 1848, was probably the first to stress the gravity of acute rheumatism in the young child, on account of the much greater liability to serious cardiac involvement. He writes: "One law respecting the connection between the cardiac and the arthritic symptoms may be stated with confidence, namely, that the younger the patient is who suffers acute rheumatism (and I have seen it so early as the third or fourth year) the more likely will he be to have rheumatic carditis I have known only three persons pass through acute rheumatism with an untouched heart prior to the age of puberty." This important observation made by Sir Thomas /

Thomas Watson nearly a century ago cannot be over-emphasised, since even today there would appear to be some who do not distinguish acute rheumatism in the child from acute rheumatism in the adult, and fail to realise the much greater havoc wrought by the disease in young children.

Garrod, in 1890, appears to have been one of the first to emphasise the protean nature of rheumatic infection and, by so doing, added greatly both to our understanding of the disease and also to the complexity of the problem. As a result of Garrod's observations, apparently trivial conditions such as tonsillitis, muscular pains and certain erythemata assumed sinister proportions when it was realised that these might signify grave danger to the cardiac muscle, a state of affairs which had hitherto been associated only with acute arthritis and chorea.

The beginning of this century was noteworthy in that Poynton and Paine, in 1900, first described a diplococcus, which they claimed to have isolated from various rheumatic lesions and which they named the diplococcus rheumaticus. While no doubt has ever been cast upon the carefulness and thoroughness of the /

the work of these observers, it is somewhat perplexing to find that subsequent investigators have failed to substantiate the claims of Poynton and Paine, and the diplococcus rheumaticus remains as elusive today as it did at the beginning of the century. Perhaps the most interesting work in connection with acute rheumatism during the past 25 years has been the attempt made to approach the disease from sociological and preventive angles and, while many such attempts have proved somewhat abortive and enthusiasm decidedly sporadic, slow but definite advance has been made, and growing interest in these aspects of the problem is apparent in many quarters today. In 1912, Poynton put forward a plea for facilities which would allow of prolonged, supervised convalescence for rheumatic children, but the Great War delayed any such steps being taken in this country until 1919, when the Invalid Children's Aid Society established a special convalescent home for rheumatic children at Willesden. The credit for instituting the first real convalescent scheme, however, belongs to America, since W. St. Lawrence, in 1915, opened a special clinic to provide for the continuous supervision and study of rheumatic children at St. /

St. Luke's Hospital, New York. This movement has shown a fairly encouraging spread during recent years with the result that many of the large cities now provide special accommodation and other facilities for the more adequate care and supervision of the rheumatic child. It would seem that development along these lines, whereby institutions are provided to cater for the very special needs and for the intensive study of rheumatic infection in children, offers the most hopeful prospect at the present time of mitigating serious cardiac disability in young people, and a concerted effort should be made in all large centres to encourage such projects.

In the following pages are detailed the results of a comprehensive clinical investigation of various aspects of acute rheumatic disease, as it is manifested in the child. Investigations of a similar nature have, of course, been carried out both in this country and abroad but, judging from the literature, no work of any magnitude dealing exclusively with juvenile /

juvenile rheumatism in the Edinburgh area has been published during the present century. The problem is so vast and has so many ramifications that it is beyond the scope of one individual to propound with any degree of authority many important factors which have a bearing on the disease, or to attempt to elucidate many of the mysteries which surround it. Details of bacteriology and immunology, questions of subtle alterations in the body chemistry, and the like, while being of great interest to the clinician are, to a large extent, outwith his sphere by virtue of the specialised knowledge and intricate technique required. The basis of this thesis is, therefore, essentially clinical and no attempt has been made to delve into the abstrusities of bacteriology and biochemistry, important as these sciences may ultimately prove to be in the solving of the problem of acute rheumatism.

The method employed comprises, in the first place, a presentation of certain general observations on acute juvenile rheumatism as it has occurred in the area served by the Royal Edinburgh Hospital for Sick Children during a period of fifteen years. Included /

Included in this section are data concerning sex and age distribution, seasonal incidence and other matters of general interest and, in addition to these, a number of clinical observations are recorded in statistical form and their significance in the scheme of acute juvenile rheumatism is discussed. By this means it is hoped to convey a reasonably accurate picture of the important types of acute rheumatic disease exemplified in children from early childhood up to the age of twelve years and, in this connection, it is necessary to define what is meant by acute rheumatic disease. In short, three manifestations are included under this heading, namely, chorea, arthritis and carditis, since there can be no real doubt that each serves as an indication of an active or recently active disease process. It may be objected that there are other rheumatic manifestations in children such as the familiar muscular pains, tonsillitis and various skin eruptions. In point of fact, the importance of these complaints is emphasised in the text and certain suggestions are put forward regarding their place in the scheme of juvenile rheumatism although, in the light of present knowledge, their inclusion in a series of acute cases is /

is scarcely justified. Further, since it is comparatively rare for a child to be brought to hospital solely on account of growing pains or occasional sore throat, particulars concerning these conditions can usually only be obtained from the histories of children admitted to hospital because of acute illness.

The second part of this work details the results of a follow-up of a large number of adolescents and young adults who had suffered from acute rheumatic disease during childhood and many of whom had not been seen within the precincts of the hospital for a number of years. It is felt that this aspect of clinical medicine is one of great importance and one that has been, to a large extent, neglected in the past. The tendency too frequently observed is to discharge a patient from hospital labelled either as "cured", "improved" or "in statu quo", and then to file away the case record and with it any further interest in the patient into the limbo of forgotten things. This is more apt to occur in the case of a patient who is apparently cured when discharged from the hospital and I would suggest, after considerable experience /

experience in follow-up work, that if the subsequent histories of such patients were methodically investigated, the optimism which prompted the discharge label "cured" would, not infrequently, have to be seriously modified. It is hoped, therefore, that the section in this thesis dealing with the melancholy picture of the rheumatic child after discharge from hospital will serve to emphasise the great importance of maintaining contact even when the disease process appears to have been arrested.

PART 1Clinical Study of a Series of Cases of Acute Juvenile
Rheumatism.

In this part of the work certain general facts relevant to the subject in question are presented in statistical form and the significance of various clinical observations is discussed. The cases under consideration were all originally diagnosed as undoubted cases of acute rheumatic disease and were under treatment as in-patients in the wards of the Royal Edinburgh Hospital for Sick Children at some period during the fifteen years, 1920 to 1934, inclusive.

In an analysis such as this carried out, as it were, in retrospect, there is one obvious difficulty, namely, the obtaining of reliable data. It is common experience that perusal of old case records inevitably reveals many important omissions and, not infrequently, the accuracy of certain of the statements contained in such records has to be questioned. This is not necessarily a reflection on those who are responsible for obtaining case histories or recording clinical details /

details, since in the rush and bustle of a busy hospital ward sins of omission and commission, common to all, are liable to be intensified. Furthermore, many facts which today are regarded as of significance were not, perhaps even ten years ago, thought to be specially worthy of record. These factors, then, must needs limit any analytical investigation which has to rely on the completeness and validity of old case records, and this will serve to explain any incompleteness which may be apparent in this work. To ensure accuracy the data presented have been very carefully selected, and a number of clinical observations have been rejected since their authenticity seemed doubtful. It is hoped, however, that the various points elaborated in subsequent pages will form a reasonably comprehensive survey of the important subject of acute juvenile rheumatism.

Yearly Incidence.

The series under discussion consists of 516 cases of acute rheumatic disease admitted to the wards of the Royal Edinburgh Hospital for Sick Children during the fifteen years, 1920 to 1934 inclusive. In connection with this disease as with other diseases, the question frequently arises as to whether the incidence /

incidence is increasing, decreasing or remaining stationary. A precise answer to this question would be possible if the compulsory notification of acute rheumatism were in force, but since this is not the case at the present time in this country, any figures dealing with incidence have to be regarded as merely approximate. It is probable, however, that a fairly accurate conception of any significant variation in incidence occurring in the Edinburgh area may be obtained by a knowledge of the admission rate of rheumatic cases to the children's hospital which serves that area. The fact that the hospital population is necessarily a selected one will not introduce any serious fallacy to a discourse on the incidence of acute rheumatic disease, since it is well known that this condition is comparatively rare in children of well-to-do parents, being largely confined to the hospital classes.

In Table 1 is shown the number of rheumatic cases admitted to the wards of the hospital each year during the period 1920 to 1934, and in the same Table the proportion of acute rheumatic cases to the total medical admissions for the year is also indicated. It must be emphasised that these figures represent new cases only, and do not include re-admissions of old cases, the number of which is very considerable. Study of the Table reveals appreciable variation from year /

year to year, but it will be noted that there has been a definite upward trend in the incidence of acute rheumatic disease, more especially during the latter five years. This is well demonstrated by dividing the fifteen years into three five year groups and showing the number of cases in each group. It is rather striking to observe that nearly one half of the total cases occurred during the five year period 1930 to 1934.

In order to ascertain to what extent this rise in incidence of juvenile rheumatism during recent years could be attributed to an increase in population, I consulted the Registrar General for Scotland who very kindly put at my disposal figures relating to the number of children under twelve years of age in the Edinburgh area during the fifteen years 1920 to 1934. It was revealed that the increase in population for this age group had been relatively small and not nearly sufficient to account for the marked rise in incidence of acute rheumatism. It may reasonably be stated, therefore, that acute rheumatism is now tending to occur more frequently amongst Edinburgh children than it did fifteen years ago.

<u>Year</u>	<u>Cases</u>	<u>Percentage of Medical Admissions</u>
1920 . . .	18 . . .	1.3
1921 . . .	25 . . .	1.9
1922 . . .	14 . . .	1.0
1923 . . .	34 . . .	2.6
1924 . . .	18 . . .	1.3
	---- 109	
1925 . . .	21 . . .	1.3
1926 . . .	39 . . .	2.5
1927 . . .	26 . . .	1.8
1928 . . .	41 . . .	3.4
1929 . . .	32 . . .	2.5
	---- 159	
1930 . . .	51 . . .	4.2
1931 . . .	32 . . .	2.7
1932 . . .	47 . . .	4.1
1933 . . .	56 . . .	5.0
1934 . . .	62 . . .	5.3
	---- 248	
<hr/> Total 516 <hr/> <hr/>		

Table 1. Showing the Number of New Rheumatic Cases admitted Each Year and the Proportion of Rheumatic Cases to Total Medical Admissions.

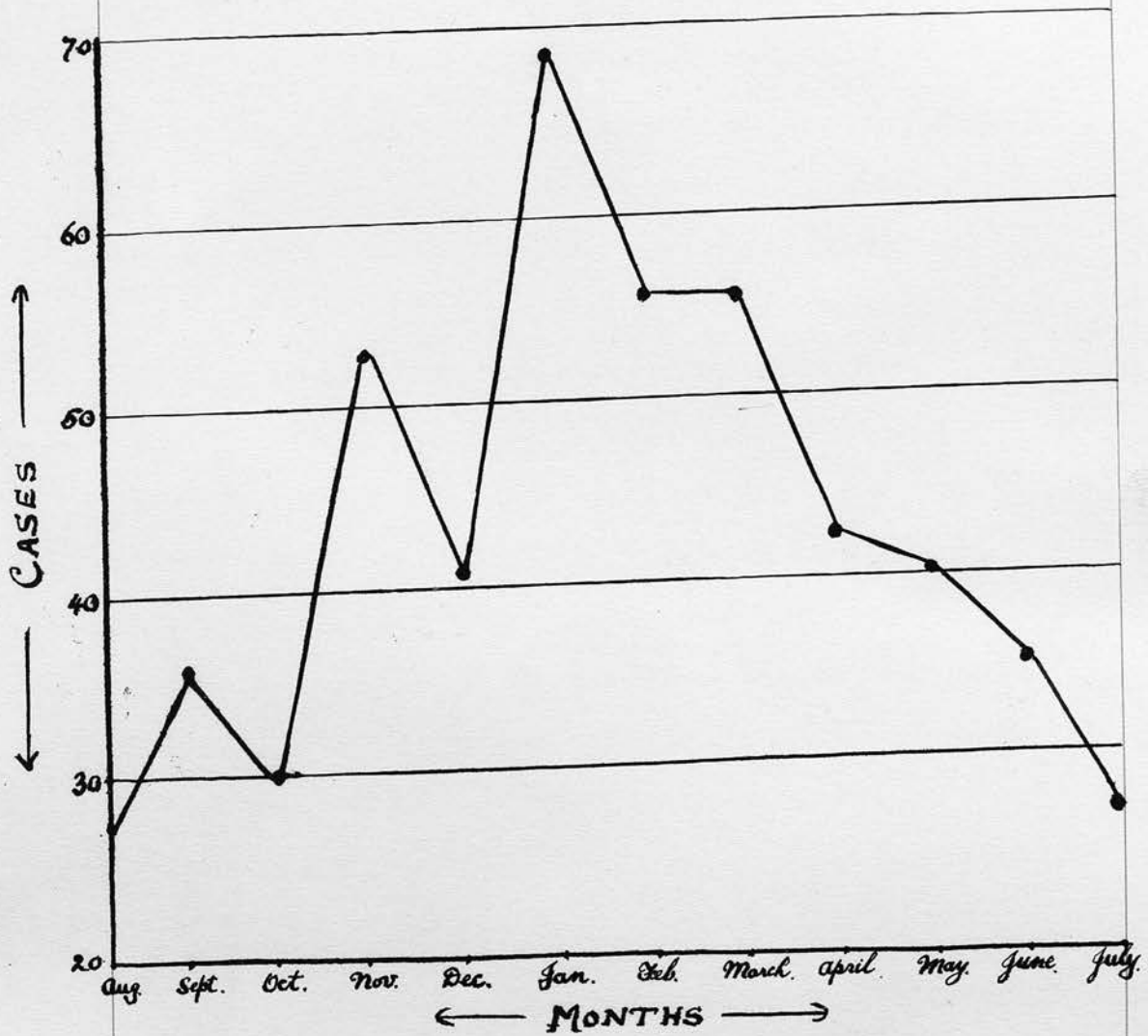
Seasonal Incidence.

It is fairly well recognised that acute rheumatism is a disease of temperate climates and that it tends to be more prevalent during the winter than during the summer months. For example, Faulkner and White (1924), working in America, found that the incidence both of rheumatic fever and chorea varied considerably with the climate and time of year, the largest number of cases occurring in the late winter and early spring. McSweeney (1931) records that 66.3 per cent. of his cases of juvenile rheumatism commenced during the cold wet months of winter and spring. On the other hand it is interesting to note that in Sydney, Australia, Maddox (1937) does not find that acute rheumatism shows any definite seasonal incidence.

In Table II is shown the monthly distribution of the cases in this series, and the figures are presented graphically in Graph 1. It will be observed that the incidence is highest during the first three months of the year, the peak month being January, and that the incidence is lowest during the high summer months of July and August. This well-defined variation between winter and summer will be referred to again in later pages in relation to possible means of preventing acute rheumatism.

<u>Month</u>	<u>No. of Cases</u>	<u>Percentage of Total</u>
January	69	13.4
February	56	10.9
March	56	10.9
April	43	8.3
May	41	7.9
June	36	7.0
July	28	5.4
August	27	5.2
September	36	7.0
October	30	5.8
November	53	10.3
December	41	7.9
<hr/> Total 516 <hr/>		

Table II. Showing the Monthly Distribution of Cases of Acute Juvenile Rheumatism.



Graph I. Showing the Monthly Distribution of
Acute Juvenile Rheumatism.

Relative Frequency of the Various Acute Rheumatic
Manifestations.

Many observers believe that acute rheumatism assumes a less virulent form today than it did 40 years ago. The view was expressed by Horder (1926) that the type of rheumatic disease had changed during the last 25 years, tending to become less severe and less acute and that the consequent cardiac lesions were more in the nature of sub-acute and chronic endocarditis rather than acute endocarditis and pericardial effusion.

It is difficult to judge, save by personal experience over a very long period, if acute rheumatism is becoming less severe in character, but it may be possible to determine whether the acute manifestations of the disease in children are undergoing any striking change in the relative frequency of their occurrence or whether the incidence of these manifestations is remaining approximately constant from year to year.

In Table III is shown the number of new cases of chorea, arthritis and primary carditis, respectively, admitted to the wards of the hospital each year during the period under consideration, and it will be observed that there has been considerable variation in the numbers /

numbers of cases of each condition from year to year. In Table IV these figures are expressed as percentages of the total for each year, thus showing the relative frequency of each acute rheumatic condition. Here again, as in the previous Table, there is a considerable annual variation, but there is nothing to suggest any significant alteration in the comparative incidence of chorea, arthritis and primary carditis in children during recent years. This is of some interest since one sometimes hears the opinion expressed that chorea is becoming relatively more common in children, a state of affairs which would be highly satisfactory. The findings in this investigation do not support this view, however, but rather indicate that chorea is merely sharing in the general rise in incidence of juvenile rheumatism as a whole.

<u>Year</u>	<u>Chorea</u>	<u>Arthritis</u>	<u>Primary Carditis</u>	<u>Total Cases</u>
1920	3	9	6	18
1921	12	9	4	25
1922	7	6	1	14
1923	20	11	3	34
1924	10	5	3	18
1925	6	10	5	21
1926	16	13	10	39
1927	12	8	6	26
1928	21	10	10	41
1929	17	5	10	32
1930	14	26	11	51
1931	14	13	5	32
1932	20	17	10	47
1933	31	21	4	56
1934	27	27	8	62
Totals	230	190	96	516

Table III. Showing the Case-Incidence of the
Three Primary Manifestations from Year to Year.

<u>Year</u>	<u>Chorea</u>	<u>Arthritis</u>	<u>Primary Carditis</u>
	Per cent.	Per cent.	Per cent.
1920	16.7	50.0	33.3
1921	48.0	36.0	16.0
1922	50.0	42.9	7.1
1923	58.8	32.4	8.8
1924	55.6	27.8	16.6
1925	28.6	47.6	23.8
1926	41.0	33.4	25.6
1927	46.2	30.7	23.0
1928	51.2	24.4	24.4
1929	53.0	15.5	31.3
1930	27.4	50.8	21.6
1931	43.8	40.7	15.6
1932	42.6	36.1	21.3
1933	55.4	37.5	7.1
1934	43.5	43.3	12.9
	44.6	36.8	18.6

Table IV. Showing the Percentage Incidence of the Three Primary Manifestations from Year to Year.

Sex Incidence

The total number of cases in this series is 516, of which 193 are males and 323 females. The male to female ratio is thus 1 to 1.7^{1.673} for the series. This general preponderance of females is not surprising since the majority of writers are agreed that acute rheumatism occurs more frequently in girls than in boys, although the actual ratio may vary considerably in different countries and even in different parts of the same country.

More detailed analysis of the sex incidence in this series of cases reveals one or two points of interest. In Table V the male to female ratio is shown for each year during the fifteen year period 1920 to 1934, and it will be noted that the ratio varies appreciably from year to year. The figures in the Table would seem to indicate, however, that in the area concerned, namely, the South East of Scotland the preponderance of females has tended to increase during recent years and, as previously mentioned, this tendency is not due to a relative increase in the frequency of chorea, as might be expected.

Table VI shows the male to female ratio at the various ages and for the purpose of comparison the ratio for the general population is given. These latter /

latter figures were obtained from the 1931 Census returns. The numbers of cases at the ages of three years and twelve years are probably too small to have any significance, but it will be observed that at the ages of ten and eleven years there is a relative increase in the number of females as compared with the other ages. It would appear, therefore, that girls, besides being more liable to develop acute rheumatism than boys, remain susceptible to the disease until they are older since, if a boy reaches the age of eleven years without having suffered from acute rheumatism, he is not then so likely to develop the disease as is a girl of the same age. Further reference will be made to this subject in the following section dealing with age distribution.

<u>Year</u>	<u>Cases</u>		<u>Ratio</u>	
	M	F	M	F
1920 -----	7	----- 11	----- 1	: 1.6
1921 -----	10	----- 15	----- 1	: 1.5
1922 -----	4	----- 10	----- 1	; 2.5
1923 -----	13	----- 21	----- 1	: 1.6
1924 -----	7	----- 11	----- 1	: 1.6
1925 -----	15	----- 6	----- 2.5	: 1
1926 -----	18	----- 21	----- 1	: 1.2
1927 -----	13	----- 13	----- 1	: 1
1928 -----	18	----- 23	----- 1	: 1.3
1929 -----	11	----- 21	----- 1	: 1.9
1930 -----	22	----- 29	----- 1	: 1.3
1931 -----	12	----- 20	----- 1	: 1.7
1932 -----	13	----- 34	----- 1	: 2.6
1933 -----	13	----- 43	----- 1	: 3.3
1934 -----	17	----- 45	----- 1	: 2.6
<hr/>				
	193	----- 323	----- 1	: 1.7
<hr/> <hr/>				

Table V. Showing the Male / Female Ratio in the Various Years.

<u>Age.</u>		<u>Cases.</u>			<u>Ratio.</u>			<u>Ratio in General Population.</u>	
		<u>M.</u>	<u>F.</u>		<u>M.</u>	<u>F.</u>		<u>M.</u>	<u>F.</u>
3	-	4	8	-	1	: 2	-	1.01	: 1
4	-	12	14	-	1	: 1.2	-	1.01	: 1
5	-	24	33	-	1	: 1.4	-	1.02	: 1
6	-	30	36	-	1	: 1.2	-	1.01	: 1
7	-	32	58	-	1	: 1.8	-	1.02	: 1
8	-	35	60	-	1	: 1.7	-	1.01	: 1
9	-	25	36	-	1	: 1.4	-	1.02	: 1
10	-	17	39	-	1	: 2.3	-	1.01	: 1
11	-	12	34	-	1	: 2.8	-	1.02	: 1
12	-	2	5	-	1	: 2.5	-	1.02	: 1

Table VI. Showing the Male / Female Ratio at the Various Ages.

Age Distribution.

Table VII shows the age distribution for the total cases in the series, and this is shown for males and females separately in Tables VIII and IX. To enable the age distribution to be appreciated more readily, Graphs II and III have been prepared. Graph II records the actual number of cases occurring at each age, whereas in Graph III the number at each age has been expressed as a percentage of the total, thus allowing of a closer comparison between males and females. It should perhaps be explained that the age distribution as shown represents the age at the first attack of acute rheumatism, so far as this could be ascertained, and not necessarily the age on admission to hospital. A proportion of the children were found to have suffered from some acute rheumatic manifestation prior to being seen at hospital, and it is the age at which the original attack commenced which is considered in this section.

It will be observed that the maximum age incidence for this series of cases is 8 years, and that this is the same for both males and females. The age distribution for this series is similar to that in the cases described by Barrett (1911), who found /

found that acute rheumatism in children was most frequent in its onset between the ages of 6 and 9 years. In the series described by McSweeney (1931) the age of onset was found to be between 5 and 10 years in the great majority of cases. Poynton and Paine (1913) discovered that in 12 per cent. of their cases the initial attack of acute rheumatism commenced before the fifth year. In the present series the corresponding figure is approximately 7 per cent.

Closer study of the figures in the accompanying Tables reveals an interesting difference between the age distributions for the two sexes since, although the largest number of cases occurs at the age of 8 years for both males and females, 35.2 per cent. of the female cases occurred after that age as compared with only 29.1 per cent. of male cases. The position is reversed at the other end of the scale, in that 46.1 per cent. of the female cases were below the age of 8 years as compared with 52.8 per cent. of the male cases. These figures lend support to the view expressed previously, that the liability to develop acute rheumatism in the later years of childhood is considerably less in boys than in girls.

<u>Age in Years</u>	<u>No. of Cases</u>	<u>Percentage</u>
3	12	2.3
4	26	5.0
5	57	11.0
6	66	12.8
7	90	17.4
8	95	18.4
9	61	11.8
10	56	10.9
11	46	8.9
12*	7*	1.4*
<hr/>		
Total	516	
<hr/>		

* Not significant.

Table VII. Showing Age Distribution for Total Cases

<u>Age in Years</u>	<u>No. of Cases</u>	<u>Percentage</u>
3	4	2.1
4	12	6.2
5	24	12.4
6	30	15.5
7	32	16.6
8	35	18.1
9	25	13.0
10	17	8.8
11	12	6.2
12*	2*	1.1*
Total		193

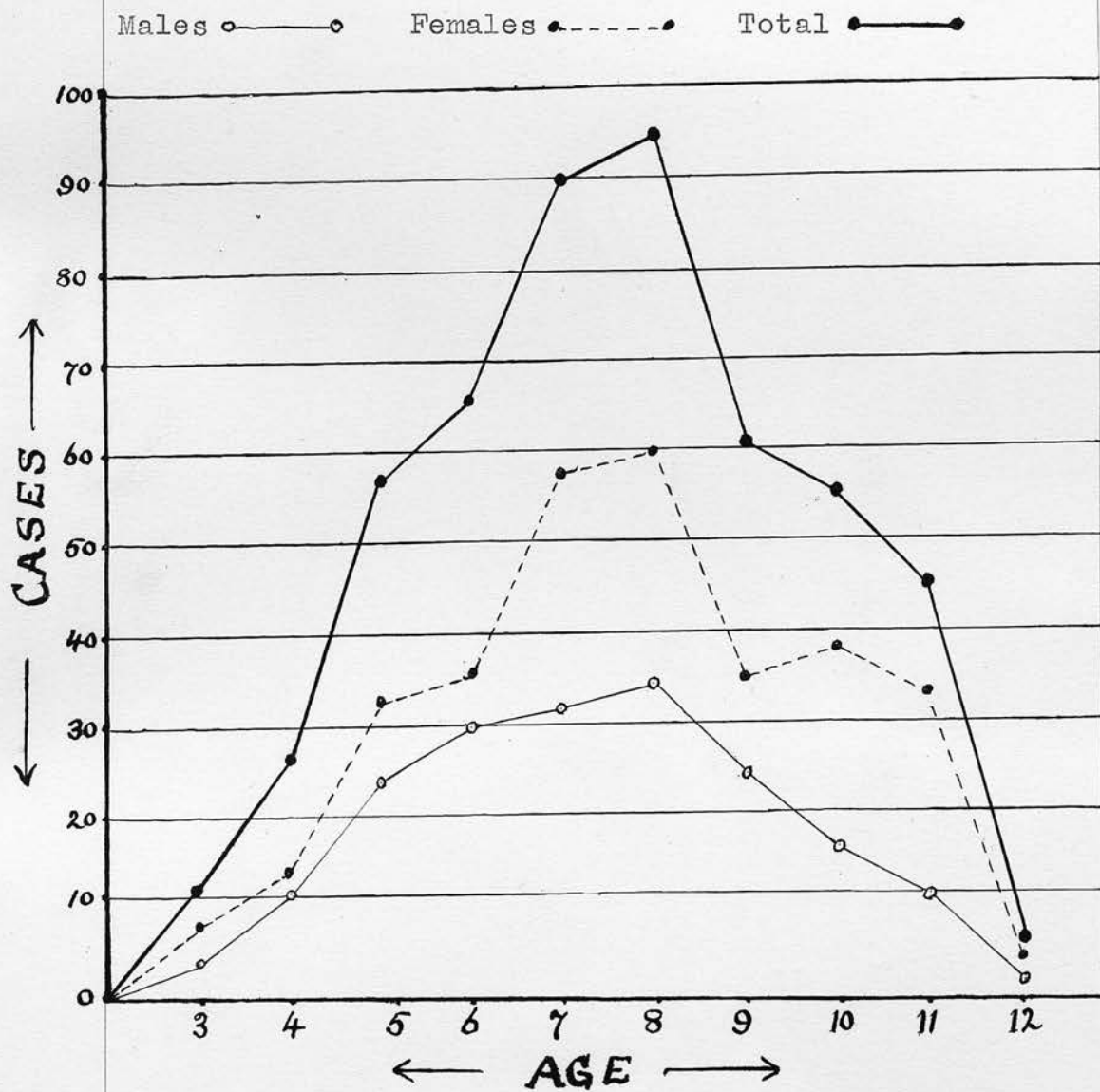
* Not significant.

Table VIII. Showing Age Distribution of the Male Cases.

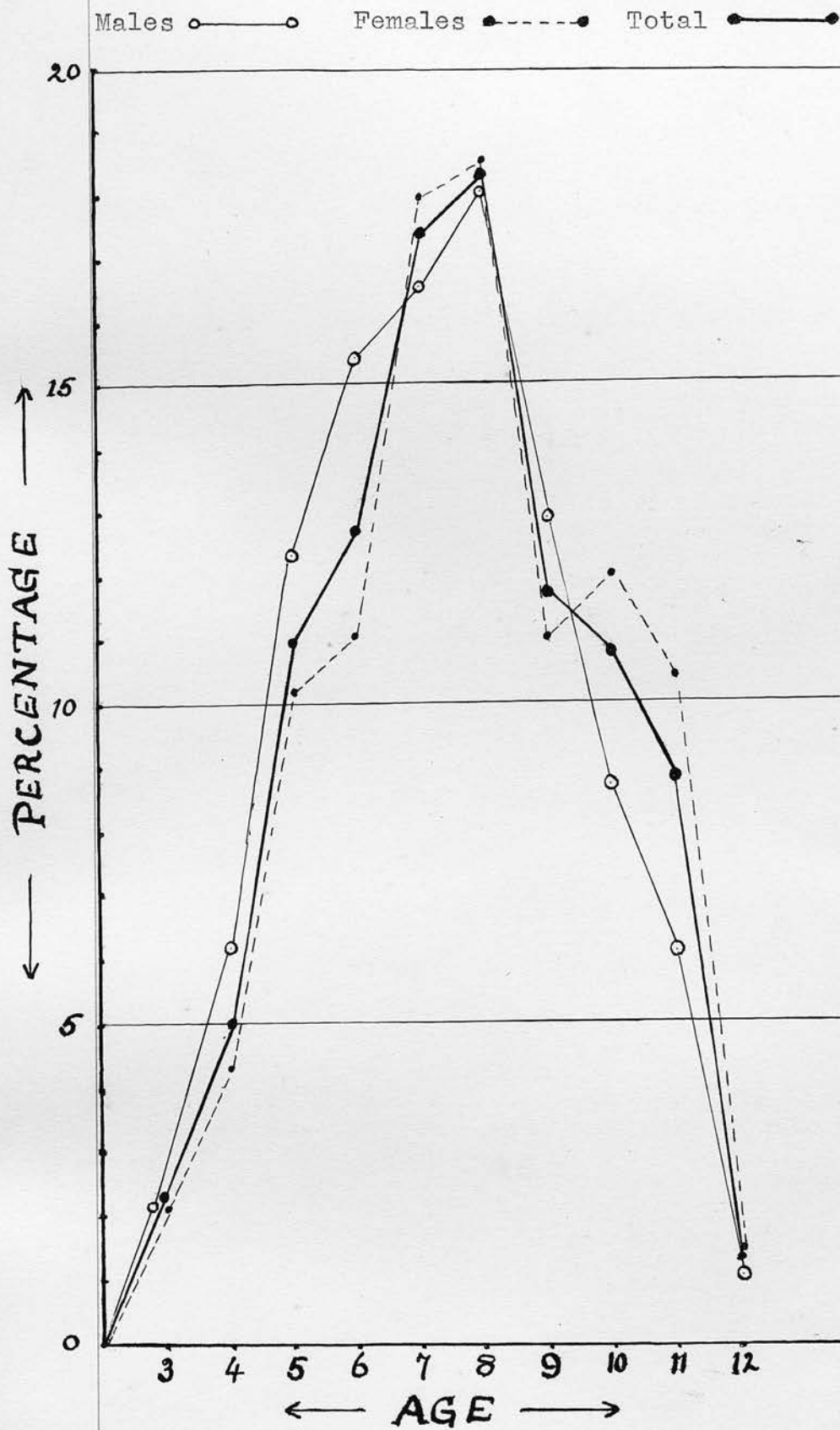
<u>Age in Years</u>	<u>No. of Cases</u>	<u>Percentage</u>
3	8	2.5
4	14	4.3
5	33	10.2
6	36	11.1
7	58	18.0
8	60	18.6
9	36	11.1
10	39	12.1
11	34	10.5
12*	5*	1.5*
<hr/>		
Total	323	
<hr/>		

* Not significant.

Table IX. Showing Age Distribution of the Female Cases.



Graph II. Showing Age Distribution of Juvenile Rheumatism.



Graph III. Showing Percentage Age Distribution to allow of Closer Comparison of Male and Female Cases.

Familial Incidence of Acute Rheumatism.

The tendency for acute rheumatism to occur in more than one member of the same family has been noted from time to time, but the reason for this is not very clear. If the disease is the result of definite infection by a specific organism then it is conceivable that a child living in a house occupied by a rheumatic subject may be infected by direct contact, more especially if conditions tend to be over-crowded. On the other hand, it is equally possible that the rheumatic constitution or tendency may be inherited by a child from its parents or grandparents, and this latter explanation would appear to be the more likely from the evidence available on the subject. It may be, however, that both factors are operative, namely, infection by direct contact and an inherited constitution. Study of the literature on the subject of familial incidence in acute rheumatism is unsatisfactory, since many authors omit to mention what they mean by family history and which members of the genealogical tree have been included in their investigations. This probably explains, to some extent, the discrepancy in the figures arrived at by various writers.

In a small series of rheumatic cases Coates and Thomas (1925) found that nearly 50 per cent gave a family /

family history of acute rheumatism. St. Lawrence (1922) working in America, reports that rheumatic children attending his clinic often had brothers and sisters with the same malady, and in 50 per cent. two or more members of the same family were affected. Thomson (1925) found a rheumatic inheritance in 25 per cent. of his cases.

With regard to the question of direct infection, it is interesting to note that Boaz and Schwartz (1927) have described two epidemics of carditis occurring at the Montefiori Hospital for rheumatic cases, and Hiller and Graef (1928) reported a similar epidemic at a convalescent home for cardiac patients. In spite of these reports, it is significant that Poynton and Schlesinger (1937) state that "on the whole there seems to be no convincing evidence, as yet, that acute rheumatism is infectious", and Miller (1926) is of the opinion that all the evidence points against the importance of direct contagion.

In this series of cases a family history of rheumatism was elicited in 155 cases, or 30 per cent. of the total, and the frequency with which the various members of the family were affected is shown in Table X. It will be observed that the scope of the enquiry was limited to the child's nearest relatives to ensure of accurate information being obtained.

In /

In Tables XI and XII the familial distribution of acute rheumatism is shown separately for males and females, and it is interesting to note that the incidence of rheumatism in the families of the male and female patients is identical, namely 30 per cent. Study of the Tables reveals one or two points of interest. It will be seen that in the majority of cases acute rheumatism had occurred in only one other member of the family besides the patient, the actual figures being 25.6 per cent. one member affected, and 4.4 per cent. two or more members affected. The incidence of acute rheumatism was found to be much higher in the mothers of the children concerned than in any other member of the family, and this is true for both boys and girls. It should perhaps be emphasised that the familial incidence of rheumatism refers to acute rheumatism only, and where there was merely a vague history of "rheumatics" affecting some member of the family this was not considered to be in any way relevant. For the purpose of control an enquiry was carried out regarding the incidence of acute rheumatism in the families of 500 children who had never suffered from rheumatic disease. It was found that in 30 of these cases, or 6 per cent., there was a definite family history of acute rheumatism. Thus it will be seen that the familial incidence is five times greater in the rheumatic group.

The /

The results of this part of the investigation supply strong evidence that acute rheumatism is, to some extent, at any rate, a familial disease and that the children of rheumatic parents, or belonging to a family in which acute rheumatism has been prevalent, run a considerably greater risk of acquiring infection than the children in whom there is not this inheritance. This fact is one that should be regarded as of major importance and reference will be made to it later in the section dealing with possible methods of preventing acute rheumatism. Further, it may be stated that the relatively high incidence of antecedent acute rheumatism in the parents of the rheumatic children supports the view that the rheumatic constitution is inherited, and that this is a factor of greater importance in the pathogenesis of the disease than infection by direct contact.

<u>Relationship</u>	<u>No. of Cases.</u>	<u>Relative Percentage.</u>
Mother	56	36.1
Father	22	14.2
Sister	20	12.9
Brother	15	9.7
Aunt	12	7.7
Grandparents	7	4.5
Father & Mother	6	3.9
Mother & Sister	5	3.2
Mother & Brother	4	2.6
Father & Sister	3	2.0
Brother & Sister	1	0.6
Two Sisters	1	0.6
Father, Mother & Sister	2	1.3
Father and two Sisters	1	0.6
<hr/>		
Total	155	(30% of total cases)
<hr/>		

Table X. Showing Details of the Familial Incidence of Acute Rheumatism in the Total Cases.

<u>Relationship.</u>	<u>No. of Cases.</u>	<u>Relative Percentage.</u>
Mother	21	36.2
Father	7	12.1
Sister	6	10.4
Brother	9	15.5
Aunt	4	6.9
Grandparents	4	6.9
Father & Mother	3	5.2
Mother & Sister	1	1.7
Mother & Brother	1	1.7
Father & Sister	1	1.7
Brother & Sister	1	1.7
	<hr/>	
Total	58	(30% of total males)
	<hr/>	

Table XI. Showing the Familial Incidence of Acute Rheumatism in the Male Cases.

<u>Relationship.</u>	<u>No. of Cases.</u>	<u>Relative Percentage.</u>
Mother	35	36.1
Father	15	15.5
Sister	14	14.4
Brother	6	6.2
Aunt	8	8.2
Grandparents	3	3.1
Father & Mother	3	3.1
Mother & Sister	4	4.1
Mother & Brother	3	3.1
Father & Sister	2	2.1
Two Sisters	1	1.0
Father, Mother & Sister	2	2.1
Father & two Sisters	1	1.0
Total		97 (30% of total females)

Table XII. Showing the Familial Incidence of Acute Rheumatism in the Female Cases.

Type of Home.

The fact that acute rheumatism is a disease largely confined to the poorer classes of the community is surely one of its most interesting aspects. This curious class distinction has been the subject of much conjecture for many years, but the problem still remains to a large extent unsolved. Naturally the questions of over-crowding and lack of adequate means of support and all that these factors bring in their train have been offered as an explanation of the prevalence of the disease amongst the poor, but, judging from the conflicting evidence which has evolved from the various investigations of this side of the problem, positive proof is extremely difficult to obtain. Not only is it difficult to assess the part played by such evil influences as over-crowding, damp and unsuitable diet in the etiology of acute rheumatism, but there appears to be considerable divergence of opinion as to whether the disease is one of extreme poverty, or whether its incidence is higher in the rather better homes of the employed working-man and artisan. Coates and Thomas (1925) believe that in the majority of cases the parents of the affected children are found to be in steady work and generally belong to the skilled artisan section of society. They believe that acute rheumatism /

rheumatism is not a disease of extreme poverty and slum dwelling. This view is supported by Thomson (1925) and by Miller (1927). Thomson states that "rheumatism is a disease of poor children, but on the whole it tends to be more frequent among the decent poor than among the squalid". In the Special Report issued by the Medical Research Council (1927), Miller endeavours to explain the higher incidence in the artisan class by pointing out that the very poor often live in an old house with stout walls, and these may keep cold and damp from the door. The artisan class live in more modern dwellings which are frequently jerry-built.

It is probably true to say that the majority of patients treated at a voluntary hospital are drawn from one of two types of home which, for descriptive purposes, may be termed slum property and artisan property. There can be no doubt that there is an increasing tendency at the present time to segregate the poorest class of patients in municipal institutions, but this applies mainly to adults. The poorest child suffering from an acute illness usually reaches the wards of the voluntary children's hospital, either by way of one of the charitable dispensaries, or brought direct by its mother. It is probable, therefore, that an analysis of acute cases admitted to a children's hospital will indicate with a fair degree of /

of accuracy the proportionate illness rate of children from slum homes and from artisan homes in the area served by the hospital in question.

In this section an attempt has been made to classify the rheumatic cases according to the type of home from which they came. The basis of classification is a broad one and is, therefore, open to considerable error, but since the control cases are classified in a similar manner it is perhaps possible to present a reasonably accurate comparison. The homes have been divided into slum and artisan, and I am indebted to the Edinburgh Public Health Authorities for kindly supplying me with the official classification of each individual house from which the town patients were drawn. This applies both to the rheumatic cases and to the control cases. A small proportion of the rheumatic cases could not be classified since they resided in districts outwith the city boundaries.

In Table XIII is shown the number of rheumatic and control cases from slum and artisan property respectively. The control cases comprise children of a similar age group admitted to the hospital suffering from some acute illness other than rheumatism. It will be noted that 15.3 per cent. of the rheumatic cases came from the poorest surroundings as compared with 12.8 per cent. of the control cases,
a /

a difference which cannot be regarded as significant. These figures are, perhaps, lower than would be expected for both groups, but they are comparable, and the surprisingly small proportion of cases from slum houses in both groups may perhaps be explained, to some extent, by an attitude of undue optimism on the part of the Public Health Authorities. After personally visiting many of the homes, I reached the very definite conclusion that the dividing line in the official mind between slum and non-slum dwellings is extremely fine in many instances.

To summarise, it may be said that in the Edinburgh area children suffering from acute rheumatism are subject to environmental influences common to the majority of children belonging to the hospital class, and although a high proportion of rheumatic children are found to live in artisan rather than slum dwellings, the same applies to children admitted to hospital with acute illness other than rheumatism.

Rheumatic Group.

	<u>Cases.</u>	<u>Percentage.</u>
Slum Property	79	15.3
Artisan Property	406	78.7
Unclassified	31	6.0

Control Group

	<u>Cases.</u>	<u>Percentage.</u>
Slum Property	99	12.8
Artisan Property	674	87.2

Table XIII. Showing a Comparison between the Type of Home from which the Rheumatic Cases came and a Control Group of Non-Rheumatic Cases.

State of Nutrition.

The impression formed as a result of examining large numbers of children suffering from rheumatic infection is that the rheumatic child tends to belong to the thin, highly-strung, asthenic type, mentally alert, but easily fatigued. It is, of course, probably true to say that a considerable proportion of all children attending hospital show some degree of inanition, but the rheumatic child is, perhaps, more consistently asthenic than other children of the hospital class. When an attempt is made to test the validity of this impression regarding the rheumatic child, one is immediately faced with the difficulty of obtaining a suitable standard for comparison. In this country there is surprisingly little authoritative information available concerning the weights and heights of children after the age of infancy, and probably the most comprehensive statistics in this connection have been compiled in America.

Graph IV depicts the weight in relation to height at the various ages of the rheumatic children in this series and, for the purpose of comparison, figures obtained from a large number of American children belonging to the poorer classes are also shown. It is apparent from the Graph that the rheumatic /

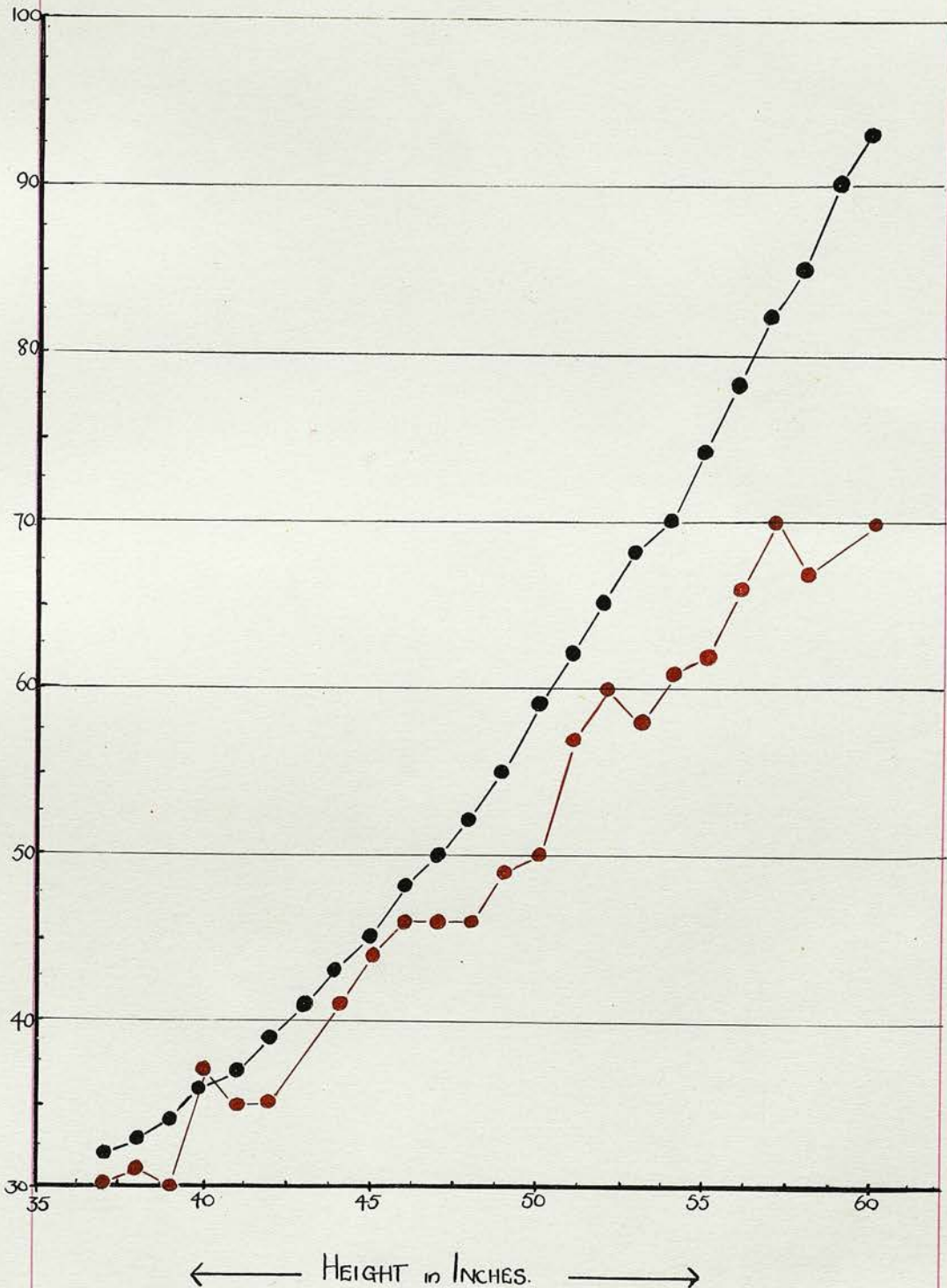
rheumatic children are consistently below the normal standard and that this deviation from normality tends to be more pronounced in the older child, reaching a maximum of 23 pounds. As has already been stated, the lack of a suitable standard for comparison is a difficulty which cannot entirely be overcome, and it is possible that British and American children are not strictly comparable. It may be stated, however, that the results of this part of the investigation lend support to the strong clinical impression that the rheumatic child is usually an undernourished child belonging to the asthenic type.

GRAPH IV.

Showing Weight in Relation to Height
between the Ages of 4 & 12 years.

Rheumatic Cases - ● -

Control Cases - ● -



First Manifestation of Acute Rheumatism.

The nature of the first manifestation of acute rheumatism cannot always be judged by the state of the child on admission to hospital since it happens,, not infrequently, that there may have been some previous attack of an entirely different character for which the child was treated elsewhere. For example, a patient admitted to hospital with chorea may, on some previous occasion, have suffered from acute arthritis or vice versa, and it is the purpose of Table XIV to give an indication of the relative frequency with which chorea, arthritis and carditis occurred as the initial manifestation of acute infection in the present series of cases.

It will be noted that the first manifestation of acute rheumatism was chorea in 44.6 per cent., arthritis in 36.8 per cent., and carditis in 18.6 per cent. It should, perhaps, be explained that the term "arthritis" does not necessarily signify an acute inflammatory condition of the joints since this is a rare occurrence in rheumatic children. In the cases classified under this heading pain in the region of the joints accompanied by pyrexia was the outstanding symptom and the one which prompted the patient to seek medical advice, although the pain in many cases was not of acute severity. The symptoms, however, were /

were of sufficient urgency to distinguish these cases from the large group in which intermittent muscular pains or growing pains were the only indications of the disease. It is significant that in nearly one fifth of the total cases the first and only signs of acute rheumatic infection were centred in the heart and, as will be seen from the Table, this does not indicate the full extent of initial cardiac damage, since in approximately one third of the chorea cases and one half of the arthritis cases, definite signs of carditis became evident in the course of the first attack although it is probable that in a fairly high proportion of these cases the cardiac damage was of a slight and transient nature. Nevertheless, in rather more than half of the cases in this series the heart was appreciably affected at the time of the original attack of acute rheumatism.

It is of interest to compare the first sign of acute rheumatism in boys and girls, since there would appear to be a definite difference in the relative frequency with which each acute manifestation occurs in the two sexes. In Table XV will be seen the incidence of chorea, arthritis and primary carditis, respectively, for boys and girls separately. It will be noted from this Table that whereas chorea is, by a fairly large margin, the commonest primary manifestation of acute rheumatism in girls, this is not so in the /



the case of boys where arthritis, by a small majority, takes pride of place. This relatively high incidence of chorea in girls has never been adequately explained and has led to an analogy being drawn between hyperthyroidism and chorea, since both conditions so markedly predominate in females. A further interesting point is the higher incidence of primary carditis which occurred in the boys of this series, since it will be seen that in approximately 26 per cent. of the male cases carditis was the first acute manifestation of rheumatism as compared with only 14 per cent. of the female cases. Furthermore, it will be observed that the incidence of initial cardiac damage in all instances is appreciably higher in the males of this series - 62.7 per cent. as compared with 46.4 per cent. of females. This difference between males and females may be explained, to some extent at any rate, by the relatively higher incidence of chorea in females, since cardiac complications are less common in chorea than in arthritis. It is, however, significant that boys appear to be more prone to develop primary rheumatic carditis than girls and this observation suggests that, although boys may be less susceptible to rheumatic infection as a whole, if they have the misfortune to contract the disease, it is liable to be of a more severe and dangerous character in that the heart will tend to be more frequently affected.

<u>First Manifestation.</u>	<u>Cases.</u>	<u>Percentage of Total.</u>
Chorea	230 (77 with carditis)	44.6
Arthritis	190 (99 with carditis)	36.8
Primary Carditis	96	18.6
	—	
Total	516	
	==	

Heart involved in 52.7 per cent. at first attack.

Table XIV. Showing the Relative Frequency of Chorea, Arthritis and Primary Carditis as the First Manifestation of Acute Rheumatism.

MALES.

<u>First Manifestation</u>	<u>Cases.</u>	<u>Percentage of Total.</u>
Chorea	64 (24 c carditis)	33.2
Arthritis	79 (47 c carditis)	40.9
Primary Carditis	50	25.9
	<hr/>	
Total	193	
	<hr/>	

Heart involved in 62.7 per cent. at first attack.

FEMALES.

<u>First Manifestation</u>	<u>Cases.</u>	<u>Percentage of Total.</u>
Chorea	166 (53 c carditis)	51.4
Arthritis	111 (52 c carditis)	34.3
Primary Carditis	46	14.2
	<hr/>	
Total	323	
	<hr/>	

Heart involved in 46.4 per cent. at first attack.

Table XV. Showing the Nature of the First
Manifestation in Males and Females.

The Incidence of Muscular Pains and Sore Throats.

It is generally agreed that recurring muscular pains, or so-called growing pains, and repeated attacks of tonsillitis in a child are in many cases indications of rheumatic infection. If this be correct then it is surely surprising that more has not been made of this knowledge, since these comparatively trivial complaints, occurring as they frequently do before the heart is affected, must serve as warnings of possible disaster in the future and demand that active precautions be taken in an endeavour to avert such disaster. With regard to the incidence of antecedent tonsillitis, Poynton (1925) in a large series of rheumatic cases found that 31 per cent. gave a previous history of tonsillitis. Bertram (1925) found preceding tonsillitis in 28 per cent. of his cases and Miller (1926) in 33 per cent. Other writers believe that the incidence is much higher than these figures quoted, for example, Ingberman and Wilson (1924) state that in their cases they obtained a history of preceding tonsillitis in 77 per cent., and St. Lawrence (1920) obtained such a history in 73 per cent. of his cases.

The great importance of growing pains as a warning sign in the child is emphasised in the Report issued /

issued by the Ministry of Health (1927) on "Acute Rheumatism in Children in its Relation to Heart Disease". In this Report the opinion is expressed that growing pains occur at some period prior to the appearance of definite acute rheumatism in the vast majority of children. Ingberman and Wilson (1924), working in America, found that growing pains had occurred in 78 per cent. of their rheumatic cases.

In the present series of cases recurring sore throats are known to have preceded the initial attack of acute rheumatism in 27.1 per cent. of all cases, a figure which corresponds very closely with the independent findings of Poynton, Bertram and Miller as mentioned previously. A history of antecedent muscular pains was obtained in 55.6 per cent. of the total cases but it is probable that this figure errs on the low side, since omissions relating to past history are not infrequent in the case records of rheumatic children.

In order to obtain a control for this part of the investigation, I interrogated the parents of 600 children belonging to the hospital class who had reached, or nearly reached the school-leaving age and who had never suffered from acute rheumatism. In these control cases the combined incidence of recurring muscular pains and tonsillitis was only 12.6 per cent. It would seem, therefore, that the relationship/

relationship between these apparently minor ailments and acute rheumatism is a close one and to regard such ailments as fortuitous or inevitable in the life of a child is both shortsighted and dangerous.

	<u>Chorea.</u>	<u>Arthritis.</u>	<u>Carditis.</u>
Tonsillitis	28.7%	27.9%	24.8%
Muscular Pains	43.5%	46.3%	70.0%

Table XVI. Showing the Incidence of Tonsillitis and Muscular Pains.

The above Table shows the percentage incidence of antecedent tonsillitis and muscular pains in relation to the subsequent acute manifestation, and it is perhaps significant that the incidence of these conditions is considerably higher in primary carditis than in either chorea or arthritis. It will be observed that no less than 70 per cent. of the primary carditis cases had suffered previously from muscular pains and yet apparently little or no attention had been paid to these warning symptoms. Careful analysis reveals that there is no appreciable difference in the incidence of tonsillitis and muscular /

muscular pains between males and females and, therefore, separate Tables for the two sexes are not shown. Nevertheless, it is significant that in both males and females the same important fact is evident, namely, the remarkably high incidence of preceding muscular pains in cases of primary carditis as compared with the chorea and arthritis cases.

Scarlet Fever in Relation to Acute Rheumatism.

It is interesting to speculate on the possible relationship between acute rheumatism and scarlet fever. If the view be accepted that the streptococcus haemolyticus plays an important rôle in the causation of acute rheumatism; then bacteriologically these two conditions have a link in common, and there can be little doubt that clinically the resemblance may at times be striking. The question as to whether an attack of scarlet fever predisposes an individual to acute rheumatism is a difficult one to answer, since the possibility of mere coincidence has to be considered. Study of the literature on the subject reveals that McSweeney (1931) found that in 6 per cent. of his rheumatic cases the onset was directly /

directly post-scarlatinal, and Bland and Jones (1935) believe that scarlet fever occurs frequently as a preceding factor in acute rheumatism, but they omit to quote actual figures.

In this series of cases the first attack of acute rheumatism immediately followed scarlet fever in 6.2 per cent., an incidence which is almost identical with that obtained by McSweeney. While it must be conceded that this figure is far from being imposing, it should be remembered that other infections did not appear to predispose to acute rheumatism. A high proportion of the children had suffered from severe illness such as measles, whooping-cough or pneumonia which, by lowering the general resistance, might have rendered them more susceptible to acute rheumatism; yet apparently this did not occur. It is at least suggestive, therefore, that scarlet fever appears in some cases to precipitate the onset of acute rheumatism, and this would lend support to the view that there exists some relationship and similarity between the two infections. Relative to this it is significant that Atwater (1927) found a definite correlation in the yearly incidence of scarlet fever and acute rheumatism in America and Wallace and Smith (1934) showed that the incidence of acute rheumatism in Edinburgh rose sharply during large epidemics of scarlet fever.

Condition of the Tonsils.

The faucial tonsils have long been regarded as an important focus of infection in rheumatic disease, so much so that the operation of tonsillectomy has become in some quarters almost a routine procedure in children suffering from acute rheumatism. The incidence of tonsillar disease in acute rheumatism, as reported by various authorities, shows remarkable variation. Lambert (1920) found infected tonsils in 25.3 per cent. of his rheumatic cases as compared with 17 per cent. in the controls. Mackie (1926) claims to have found infected tonsils in 58.8 per cent. of his rheumatic patients as compared with 27.5 per cent. of his control cases. Miller (1926) states that 83 per cent. of his cases showed diseased tonsils, but he omits to mention the important question of controls.

Table XVII shows the condition of the tonsils in the children in this series as recorded at the first examination of the throat. It is fully appreciated that the appearance of tonsils on examination is apt to be somewhat deceptive and, even amongst experts, opinions differ as to when, for instance, the tonsils are merely hypertrophied and when they are actually the seat of infection. The figures shown in this Table, therefore, must be treated with some reserve /

reserve, but they will give a general indication of the state of the throat in the cases under consideration. It will be noted that at the time when these cases were first examined the tonsils were considered to be diseased in 161 cases, or 31.2 per cent., and tonsillectomy had been performed prior to the onset of acute rheumatism in a further 67 cases, or 13 per cent. It has to be presumed that in these latter cases the tonsils must have been considered unhealthy otherwise they would not have been removed, and the total incidence of tonsillar disease was thus 44.2 per cent. In a control series of non-rheumatic children the total incidence of tonsillar disease was found to be somewhat less than in the rheumatic cases, namely, 30 per cent., and, as mentioned in a previous section, the incidence of sore throat was much lower in the control cases than in the rheumatic cases.

	<u>Healthy</u>	<u>Enlarged</u>	<u>Diseased</u>	<u>Removed</u>	<u>Totals</u>
Chorea —	87	43	65	35	230
Arthritis —	79	28	61	22	190
Carditis —	37	14	35	10	96
<u>Totals</u> —	203	85	161	67	516

Table XVII. Showing Condition of Tonsils at First Examination.

Tonsillectomy in Acute Rheumatism.

A question which constantly arises in connection with the complex problem of juvenile rheumatism is the advisability of tonsillectomy as a prophylactic measure and the fact that great divergence of opinion is apparent in the literature on this subject merely serves to emphasise that, as yet, no finality has been reached on this important matter. Starling (1923) states that "enucleation of tonsils is a most valuable means by which the acute attack may be ended and further infection prevented. The earlier the tonsils are removed the greater is the benefit derived". Hunt and Osman (1923) reporting on a series of rheumatic children, observe that in those who had had their tonsils removed recurrences occurred in 53 per cent., and where no operation had been performed the recurrence rate was only 42 per cent. Robey and Freedman (1927) believe that enucleation of the tonsils offers the best preventative of rheumatic fever and, therefore, of rheumatic heart disease, whereas Farnum (1928) is of the opinion that removal of tonsils will not prevent spread of infection. Wilson, Lingg and Croxford (1928) found that as a prevention against a rheumatic attack the results of this operation were discouraging. Poynton and Schlesinger (1937) sum up /

up the situation by stating that "there is sufficient proof of the intimate association of throat infection and rheumatism to warrant removal of the tonsils when they are septic, but until there is decided evidence that children without tonsils are definitely protected against rheumatism, there appears to be no justification for the operation when the throat is healthy".

If the tonsils are regarded as being an important focus of infection, then it would seem reasonable to suppose that, if this focus were removed in early life, the child would be less liable to develop acute rheumatism at a later date, and in order to investigate this important matter I enlisted the expert collaboration of Dr. A. Brownlie Smith.

It is well recognised that acute rheumatism, although it may occur, is comparatively uncommon before the age of five years, and the scheme adopted was to obtain from the hospital records names of children in whom tonsillectomy had been performed before the age of five years and who had not suffered previously from any rheumatic manifestation. These children, numbering in all 403, were requested to report for questioning and for examination of the throat and at the time of reporting they had all reached, or were approaching, the school-leaving age.

It /

It was thus possible to obtain information regarding their health throughout the whole period of childhood.

In order to secure the necessary controls, we obtained from the hospital records the names of children who had had their tonsils removed during the later years of childhood. A total of 574 children in this category were interrogated regarding the incidence of acute rheumatism, and the results obtained were compared with the other group. It would have been interesting also to have had a series of cases in which no operation had been performed on the throat but, unfortunately, it was not possible to obtain a sufficient number of such cases to warrant their inclusion here for the purpose of comparison and control. To ensure that the information elicited was reliable and definite, the title "acute rheumatism" was restricted to three manifestations, namely, chorea, acute arthritis and rheumatic carditis, while the less acute conditions such as muscular pains and sore throats were not considered.

A factor which had to be taken into account in an investigation of this nature was the completeness of the tonsil operation and special care was taken, therefore, in the examination of the throats of the children who reported. In the first group of children /

children in whom the tonsils had been removed before the age of five years, the incidence of acute rheumatism during the interval between the removal of tonsils and the school-leaving age was 7.2 per cent., whereas in the control group the incidence of acute rheumatism was 4.2 per cent. In Table XVIII an analysis of the frequency of the various rheumatic manifestations is shown for the two groups.

The results obtained from this part of the investigation would appear to afford strong evidence that removal of the tonsils in early life does not lower the incidence of subsequent rheumatic infection and that this operation, performed at an age when it might be expected to be of considerable benefit, not only fails completely to protect the child against acute rheumatism, but may even render him more liable to develop the disease. It would seem, therefore, that in the absence of unequivocal signs of local sepsis the most reliable indication for tonsillectomy in a child is the occurrence of repeated attacks of sore throat and, in the absence of these, the operation is probably of little value in controlling acute juvenile rheumatism, and may even be fraught with considerable risk.

TONSILLECTOMY GROUP.

	<u>Cases.</u>	<u>Percentage.</u>
No Rheumatic Condition	374	92.8
Chorea	6	1.5
Acute Arthritis	16	4.0
Chorea & Carditis	2	0.5
Arthritis & Carditis	4	1.0
Chorea & Arthritis	1	0.2
<u>Total</u>	<u>403</u>	

CONTROL GROUP.

	<u>Cases.</u>	<u>Percentage.</u>
No Rheumatic Condition	550	95.8
Chorea	7	1.2
Acute Arthritis	6	1.0
Primary Carditis	4	0.7
Chorea & Carditis	3	0.5
Arthritis & Carditis	2	0.4
Chorea & Arthritis	2	0.4
<u>Total</u>	<u>574</u>	

Table XVIII. Showing Details of the Rheumatic Manifestations which occurred in the Tonsillectomy Group and the Control Group.

Rheumatic Nodules.

It is extremely difficult to state with any degree of accuracy the frequency with which subcutaneous nodules occur in a large series of rheumatic children. This is due to the fact that such lesions are notoriously transitory in their appearance and it is thus highly probable that their presence is frequently overlooked. In a condition such as acute rheumatism in which recurrences are so frequent, it is common experience to note the presence of nodules during one acute phase of the illness and not during another, and unless rheumatic children are kept under constant supervision for a period of years, any attempt to estimate the true incidence of rheumatic nodules can but be very approximate. This difficulty probably accounts for the great diversity of views expressed in the literature regarding the frequency with which nodules occur. For example, Maddox (1937) reports the presence of nodules in only 1.5 per cent. of a large group of rheumatic children in Sydney. Bertram (1925) found nodules in 7 per cent. of his cases in Glasgow, whereas Benjamin (1927) records an incidence of 15.6 per cent.

Although no claim is made to accuracy, it may be of interest to allude to the recorded incidence of rheumatic /

rheumatic nodules in this series of cases. Out of a total of 516 cases nodules were observed in only 47, which represents 9.1 per cent. of the total. This low incidence is, I suggest, significant in that it illustrates the elusive character of these rheumatic lesions, since they probably occurred with much greater frequency than this figure suggests. The incidence of nodules was found to be slightly higher in males than in females - 10.3 per cent. and 8.8 per cent. As would be expected the great majority of cases in which nodules were noted were suffering from carditis. Of the 47 cases, 42 or 89 per cent. were found to have some cardiac defect, and in the remaining 5 cases the heart was apparently unaffected. In 29 instances the subcutaneous nodules were reported as being numerous and were found to occur in various parts of the body, whereas in the remaining 18 cases the nodules were confined to the region of one joint. It is of interest to note that where the nodules were monarticular the elbow was the joint affected in almost every case, the left more frequently than the right.

The Importance of Epistaxis as a Symptom.

It is perhaps strange that so little emphasis is given to the association of the symptom of epistaxis with acute rheumatic infection in childhood. Brief reference is made to its occurrence in text-book descriptions of the disease, but it is rarely stressed as a symptom of importance. The causation of severe epistaxis in acute rheumatism is somewhat obscure. Coburn (1933) believes that the early inflammatory tissue changes in rheumatic infection are accompanied by an engorgement of blood vessels and an increased permeability of their walls, giving rise to a general haemorrhagic tendency during the active stages of the disease. If this belief be correct then, presumably, epistaxis may be regarded as an indication of this haemorrhagic tissue change.

In the series of cases under discussion the frequency with which nose-bleeding occurred was ascertained in 227 cases, and in the remainder the evidence was not considered to be sufficiently reliable to warrant a definite opinion. Severe, repeated attacks of epistaxis occurred in 68 of the 227 cases, which represents 30 per cent. It was learned that in a number of cases slight, transient haemorrhage had occurred, but these have been disregarded as being probably fortuitous and of no significance, and only those cases in which the symptom was severe and recurring /

recurring have been considered. More detailed analysis of the incidence of epistaxis reveals the fact that it was essentially a symptom of active rheumatism and that it was frequently associated with cardiac changes, since in no less than 78 per cent. of the cases in which it occurred active carditis was present, as compared with an incidence of only 22 per cent. in cases where the heart was not involved. For the purpose of a control to this part of the investigation, 300 adolescents who had never suffered from acute rheumatism during childhood were questioned regarding the occurrence of epistaxis, and it was found that only 1.2 per cent. had suffered from severe haemorrhage.

Thus it would seem that severe, repeated epistaxis is a symptom which accompanies acute juvenile rheumatism with relative frequency and, as such, it should be regarded as of considerable importance in the symptomatology of the disease. Furthermore, it is a symptom which appears to be specially associated with the active phase of the disease and the sudden occurrence of severe nose-bleeding in a case which is thought to be quiescent should rouse strong suspicions of imminent recrudescence.

Rheumatic Pneumonia.

The occasional association of pulmonary complications with acute rheumatism in childhood has been noted by several authors, but the nature and significance of such complications remain somewhat obscure. The interesting question arises as to whether there is such a condition as rheumatic pneumonia due to an actual invasion of the alveolar tissue by the rheumatic process, or whether the occurrence of pneumonic consolidation in the course of acute rheumatism is merely incidental and not in any way a specific infection. Riesman (1921) reports finding signs of pneumonic consolidation of the left lung occurring in pericarditis, but believes that this is not true pneumonia but due to compression of the lung by the heart. Naish (1928) studied consolidation of the lungs in six fatal cases of acute rheumatism. This author found an inflammatory reactive process which, he states, bore a strong resemblance to the lesions described by Tawara, Carey Coombs and Aschoff as pathognomonic of rheumatic infection in other parts of the body. The appearance was quite unlike that seen in other forms of pneumonia and Naish does not subscribe to the view that the condition is due to collapse from pressure of an enlarged heart.

The /

The incidence of pneumonic consolidation of the lungs has been carefully investigated in this series of cases and it is found to have been present, without doubt, on 18 occasions. This is, admittedly, a small figure, although it actually represents 3.5 per cent. of the total. So far as could be ascertained, in all these cases there was a true pneumonic consolidation as distinct from pleural effusion or acute bronchitis. It is of interest to note that this type of pneumonia was never found in association with chorea although it did occur in the absence of active rheumatic carditis, since in 4 of the cases mentioned there was no obvious heart lesion present at the time when the pneumonia developed. In the remaining 14 cases there were definite signs present of advanced active carditis. It was determined that rheumatic consolidation of the lungs occurred in 5.2 per cent. of the boys in this series as compared with an incidence of 2.5 per cent. in the girls. This is perhaps significant, since it lends support to the view previously expressed that acute rheumatism, although not so prevalent in boys, is apt to be of a more serious character when it does occur.

These observations suggest that there may be a definite entity deserving the title of rheumatic pneumonia if it is permissible to base such a surmise on purely clinical data. It is evident that when pneumonic /

pneumonic consolidation does occur in acute rheumatism it is usually associated with severe cardiac decompensation, although it has been observed in a few cases where the heart appeared to be unaffected.

SUMMARY of PART I.

A clinical survey of 516 cases of acute juvenile rheumatism is presented. All of these had been In-Patients at the Royal Edinburgh Hospital for Sick Children during the fifteen year period 1920 to 1934 inclusive.

The incidence of the disease showed a definite tendency to rise during the period under consideration and it was ascertained that this rise was not due to an increase in population.

The seasonal variation of acute rheumatism is shown and it is noted that the disease is more prevalent during the winter months, the largest number of cases occurring during the first quarter of the year.

The male to female ratio for this series of cases is 1 to 1 .7 and it is revealed that female preponderance has tended to increase during recent years. It is further noted that this preponderance becomes more evident during the later years of childhood, thus suggesting that girls are more liable than boys to develop acute rheumatism at the somewhat critical period which precedes puberty.

The age distribution is discussed and it is shown that the first attack of acute rheumatism occurred most frequently at the age of 8 years in both sexes.
In /

In 7 per cent. of the total series the acute symptoms commenced before the age of 5 years.

The relative frequency of chorea, arthritis and primary carditis as the first manifestation of acute rheumatism in this series was ascertained. It is shown that, although there is considerable variation from year to year, no significant alteration in the relative incidence of these three manifestations has occurred during recent years.

The high incidence of cardiac damage in the course of the first attack of acute rheumatism is stressed, and it is revealed that the incidence is appreciably higher in boys than in girls.

The familial incidence of the disease is discussed. In this series, enquiry elicited the information that a definite family history of acute rheumatism existed in 30 per cent. of the cases. This figure becomes very significant when compared with a large series of control cases where the familial incidence of acute rheumatism was only 6 per cent.

The homes from which the rheumatic children came were investigated and it was revealed that only a small proportion of these children occupied slum property. When compared with a series of control cases, however, no significant difference in home conditions was discovered between the rheumatic children and /

and other children belonging to the hospital classes.

The state of nutrition of the rheumatic children as evinced by weight in relation to height was investigated, but difficulty was experienced in obtaining a suitable standard with which to compare them. It may be stated, however, that the available evidence lends support to the clinical impression that the rheumatic child is an under-nourished child belonging to the asthenic type.

The frequency and importance of antecedent muscular pains and sore throats is discussed, and their significance as warning signs is emphasised.

The frequency with which scarlet fever immediately preceded the onset of acute rheumatism is shown, and it is suggested that there is possibly a similarity and relationship between the two diseases.

The condition of the faucial tonsils at the time of the first examination of the throat is described, and the advisability of tonsillectomy as a prophylactic measure in acute rheumatism is carefully considered.

The significance of severe epistaxis as a frequent symptom of acute rheumatism in children is discussed and the value of this symptom as an indication of active disease is noted.

The incidence of subcutaneous nodules in this series /

series of cases is discussed and reference is made to their distribution.

The occurrence of rheumatic pneumonia in this series is considered and it is revealed that, in those cases where associated consolidation of the lungs did occur, boys were more frequently affected than girls.

PART II.The Later Effects of Acute Juvenile Rheumatism.

The study of the young child during an attack of acute rheumatism is, admittedly, of great importance, but it is an incomplete picture and one that fails to portray the real gravity of the problem. In cases of primary carditis where the patient is discharged from hospital with signs of permanent cardiac damage, the ultimate catastrophe can be foretold with comparative certainty, although even in such cases the unexpected may occur as will be shown in later pages. It is in the less severe manifestations of rheumatism - chorea and arthritis - where the immediate results of the acute illness may be totally misleading, and where frequent contact with the patient following discharge from hospital is of paramount importance. This becomes evident when it is recalled that one of the most interesting facts concerning acute rheumatism is its liability to recur more than once in the same individual, thus distinguishing it from other infections where one attack has the effect of producing immunity against subsequent attacks. This tendency to acute relapses is /

is one of the most disquieting features about the disease, since it renders an accurate prognosis well nigh impossible. A feature about this tendency to recurrence, which is well shown in subsequent pages, is the fact that such recurrences are not, so to speak, true to type and it is quite impossible when attempting to forecast the course of events in, for example, a case of chorea, to express a definite opinion as to whether any relapses are likely to occur at all and, if they do occur, what form they are likely to take. If acute recurrences were always similar in character to the initial attack then the position would be simplified, but this is far from being the case as will be shown.

In this section it is hoped to depict the plight of a number of adolescents and young adults who were the victims of acute rheumatic disease during childhood. When an attempt is made to follow up patients after the lapse of a number of years one insuperable difficulty is always encountered, namely, the locating or tracing of many of these patients. It is a frequent and disheartening experience to have many letters of enquiry "returned unknown", and the available material is thereby much reduced. Under the circumstances this unknown factor can only be disregarded altogether and this may constitute a fallacy in the interpretation of the findings in an enquiry /

enquiry of this nature, since it is quite impossible to form any precise opinion regarding the present condition of the patients untraced. It is only possible to presume that their present state of health corresponds with that of the patients about whom information is available and that, therefore, the accuracy of the findings will not be prejudiced.

The procedure adopted in this follow-up aimed at obtaining the maximum amount of clinical information relating to the present state of health of the individuals concerned, combined with accuracy of observation. For this reason the now popular questionnaire method was discarded, so far as possible, in favour of the individual examination of each patient and the careful interrogation of the patient's mother or father. In some instances circumstances did not permit of the patients coming to hospital for examination and where possible these were visited in their homes or information regarding their health was obtained from their family doctors.

For this part of the investigation the patients selected for examination had all reached adolescence or adult life, their ages ranging from 15 to 20 years, and it was thus possible to obtain a reasonably complete picture in retrospect of the nature and course of juvenile rheumatism from the time of the first attack until adolescence was reached or until death /

death supervened. The progress of these individuals during adult years is, of course, unknown since they are at present merely on the threshold of mature life, but knowledge of their condition now may serve as a pointer for the future and indicate the general trend of events in this unabating scourge of childhood.

The method employed was to write to the patients themselves, or to their parents, and request them to attend at hospital for examination. The response to this follow-up procedure was very encouraging considering the fact that a number of years had elapsed since these patients had been in contact with the hospital. In many instances it was both interesting and gratifying to note the obvious pleasure and surprise displayed by the parents at the interest which was being taken in their children, but it seemed also that their very apparent surprise was by way of being a reproach on the indifference and neglect which is unfortunately so characteristic of many where the rheumatic child is concerned.

It is proposed in this section to discuss separately the sequence of events in chorea, arthritis and primary carditis, respectively, and then briefly to summarise the later effects of acute juvenile rheumatism as it occurs in the area served by the Royal Edinburgh Hospital for Sick Children.

Later Effects of Chorea.

Detailed information regarding the state of their health in adolescence was determined in 135 cases of juvenile chorea, and the findings may be presented as follows.

Recurrences.

In this series, 63 per cent. of the children who had suffered from chorea as the first manifestation of acute rheumatism were subject to relapses and a high proportion of these suffered from more than one relapse. The sequence of events from the initial attack of chorea until adolescence was reached is detailed in Table XIX.

It will be observed that 50 cases did not relapse after the initial attack of chorea; in 33 cases only one recurrence occurred, whereas in the remaining 52 cases (38.5 per cent.) two or more recurrences are known to have occurred in the interval between the first attack of chorea and adolescence. The variation in type of relapse will further be noted, although the general tendency is for chorea to follow chorea.

Condition on Examination.

With regard to the actual physical condition of the chorea patients, as previously stated detailed information /

information concerning 135 cases has recently been obtained and, since the whole problem of acute rheumatism is centred in the heart, the well-being of these patients was assessed in terms of cardiac efficiency. The later effects of chorea and of the other primary manifestations have thus been classified under three headings - "Heart normal"; "Organic cardiac lesion present"; "Dead". These three groups will now be considered individually for the chorea cases.

Heart Normal. Under this heading 87 cases have been included which represents 64.4 per cent. of the patients examined. After thorough examination no abnormality, either in the heart or elsewhere, could be detected in these patients and they were all considered capable of full employment and of indulgence in vigorous sports.

Organic Cardiac Lesion Present. In this group there were 30 cases representing 22.2 per cent. One half of the patients comprising this group were found to be in excellent general health despite the presence of a definite cardiac lesion, and a number of them regularly indulged in strenuous forms of exercise such as swimming and cycling without any ill-effects. The remainder of the patients included under this heading showed varying degrees of cardiac decompensation which had the effect of restricting many of the normal /

normal activities of healthy adolescence. The individual cardiac lesions discovered on examination of these 30 patients may be summarised as follows:-

Mitral Regurgitation . . 23 cases

Mitral Stenosis 6 cases

Aortic Regurgitation . . 1 case

Dead. As the result of careful enquiry it was ascertained that 18 cases of chorea out of the total of 135 had failed to survive to adolescence. This figure represents a mortality rate of 13.3 per cent., a somewhat formidable proportion when it is remembered that chorea is the least of the evils of acute juvenile rheumatism. Investigation into the cause of death in these cases revealed the fact that 15 died from rheumatic carditis, whereas in the remaining 3 cases death was due to illness unconnected with acute rheumatism. For ease of reference, the late results of chorea in childhood as described above are shown in Table XX.

Recurrences. (CHOREA)

<u>1st</u>	<u>2nd</u>	<u>3rd</u>	<u>4th</u>	<u>5th</u>	<u>No. of Cases.</u>
No Recurrences					50
C.	-	-	-	-	22
A.	-	-	-	-	3
Ca.	-	-	-	-	5
C.Ca.	-	-	-	-	2
A.Ca.	-	-	-	-	1
C.	C.	-	-	-	12
C.	C.Ca.	-	-	-	1
C.	A.	-	-	-	1
C.Ca.	C.Ca.	-	-	-	3
A.Ca.	C.	-	-	-	1
A.Ca.	Ca.	-	-	-	2
A.Ca.	A.Ca.	-	-	-	1
C.	C.	C.	-	-	9
C.	A.Ca.	A.Ca.	-	-	2
C.Ca.	C.Ca.	Ca.	-	-	1
A.	A.	A.	-	-	1
C.	C.	C.	C.	-	4
C.	C.	C.	C.Ca.	-	1
C.	C.	A.Ca.	C.Ca.	-	1
C.Ca.	C.C.	C.Ca.	C.Ca.	-	2
C.	C.	C.	C.	C.	4
C.	C.	C.	C.	A.Ca.	1
C.	C.	C.	C.Ca.	C.Ca.	1
C.	C.	C.	A.Ca.	Ca.	1
C.Ca.	C.Ca.	C.	C.	C.	1
A.	A.	A.Ca.	C.Ca.	C.Ca.	1
A.Ca.	A.Ca.	C.Ca.	C.Ca.	Ca.	1
<u>Total</u>					<u>135</u>

KEY.

C.	-	Chorea.
Ca.	-	Carditis.
A.	-	Arthritis.
C.Ca.	-	Chorea with Carditis.
A.Ca.	-	Arthritis with Carditis.

Table XIX. Showing the Sequence of Recurrences in the Chorea Cases.

CHOREA.

	<u>Cases.</u>	<u>Percentage.</u>
Normal	87	64.4
Mitral Regurgitation	23	17.0
Mitral Stenosis	6	4.4
Aortic Regurgitation	1	0.8
Dead	18	13.3*
<u>Total</u>	<u>135</u>	

* Corrected Mortality - 11.1 per cent.

Table XX. Showing the Present Condition of the
Chorea Cases.

Later Effects of Acute Arthritis.

In 124 cases where arthritis was the first manifestation of acute rheumatism detailed information is available regarding their progress from the time of the first attack until adolescence, and the data obtained are recorded below.

Recurrences.

In this series of arthritis cases the recurrence rate was very high, 71 per cent. of the patients having suffered from subsequent attacks. As was the case in chorea, many of the patients had more than one acute relapse and the nature of the sequence of these relapses is shown in Table XXI. It will be seen from this Table that multiple relapses occurred in 39 of the arthritis cases, or 31.5 per cent. of the total. In this respect comparison with chorea is of interest since it transpires that, whereas the actual recurrence rate in arthritis is higher than that of chorea, the proportion of multiple relapses is slightly greater in chorea.

Condition on Examination.

Detailed information was obtained regarding the present condition of 124 cases of arthritis and this may be conveniently related as follows.

Heart Normal. On examination 45 patients were found to /

to have hearts which were normal in every respect, and the state of the general health corresponded with this finding in that the individuals concerned were able to work and play without exhibiting any undue signs of fatigue.

Organic Cardiac Lesion Present. Under this heading 42 patients were included and of these 27 were found to be enjoying good health and regular employment despite the presence of undoubted cardiac damage. The remainder exhibited definite signs of heart failure which necessitated the curtailment of normal activities to a varying extent and compelled many of them to lead very restricted lives.

The cardiac lesions discovered when these patients were examined were as follows:-

Mitral Regurgitation . . 36 cases

Mitral Stenosis 4 cases

Aortic Regurgitation . . 2 cases

Dead. It was learned that, out of the total of 124 arthritis patients about whom complete data was recently obtained, 37 had died before the age of puberty representing a mortality of 29.8 per cent. Of these, 35 died as a result of rheumatic carditis and 2 from other causes. These figures can but emphasise the gravity of the situation when a young child develops joint symptoms which in themselves may be comparatively slight.

The later effects of arthritis in childhood are summarised in Table XXII.

Recurrences. (ARTHRITIS)

<u>1st</u>	<u>2nd</u>	<u>3rd</u>	<u>4th</u>	<u>5th</u>	<u>No. of Cases.</u>
No Recurrences					36
C.	-	-	-	-	16
A.	-	-	-	-	10
Ca.	-	-	-	-	14
A.Ca.	-	-	-	-	7
C.Ca.	-	-	-	-	2
C.	C.	-	-	-	3
C.	A.	-	-	-	2
C.	Ca.	-	-	-	1
A.	C.	-	-	-	3
A.	A.	-	-	-	1
A.	Ca.	-	-	-	2
A.Ca	Ca.	-	-	-	6
Ca.	Ca.	-	-	-	2
C.Ca.	C.Ca.	-	-	-	3
A.Ca.	C.Ca.	-	-	-	1
C.	C.	C.	-	-	1
C.	C.	Ca.	-	-	1
A.	A.	A.	-	-	2
A.	C.	C.	-	-	1
A.	A.	A.Ca.	-	-	2
A.	A.Ca.	Ca.	-	-	1
A.Ca.	A.Ca.	Ca.	-	-	3
Ca.	Ca.	A.Ca.	-	-	1
C.	C.	C.	C.	-	1
C.	C.	C.Ca.	C.Ca.	-	1
A.	A.	A.	A.Ca.	A.Ca.	1
<u>Total</u>					<u>124</u>

KEY.

C.	-	Chorea.
A.	-	Arthritis.
Ca.	-	Carditis.
C.Ca.	-	Chorea with Carditis.
A.Ca.	-	Arthritis with Carditis.

TABLE XXI. Showing the Sequence of Recurrences in
the Arthritis Cases.

ARTHRITIS.

	<u>Cases.</u>	<u>Percentage.</u>
Normal	45	36.3
Mitral Regurgitation	36	29.0
Mitral Stenosis	4	3.2
Aortic Regurgitation	2	1.6
Dead	37	29.8*
<u>Total</u>	<u>124</u>	

*Corrected Mortality - 28.2 per cent.

TABLE XXII. Showing the Present Condition of the Arthritis Cases.

Later Effects of Primary Carditis.

The aftermath of primary rheumatic carditis in children is truly a melancholy picture. At once the most insidious and the most deadly of all manifestations, it strikes with little or no warning and exacts a heavy toll of young lives, or leaves its victims permanently crippled to eke out a burdensome existence devoid of all hope.

In the present series reasonably comprehensive information regarding the late-effects of primary carditis was obtained in 66 cases. In many instances the patients in this group were too ill to attend hospital for examination but this difficulty was overcome, whenever possible, by visiting the homes of the patients who still survived and interviewing the parents of those children who had died in the interval.

Recurrences.

The frequency of relapses in the cases of primary carditis was found to be comparatively low when compared with the frequency in chorea and arthritis. Of the 66 patients regarding whom information is available, only 16 or 24.2 per cent. were found to have suffered from acute recurrences. This low recurrence rate in comparison with the rest of the series is due in part to the fact that a high proportion /

proportion of the children who developed primary rheumatic carditis died in the course of the initial attack. The recurrence rate in those cases which survived the initial attack was 45.7 per cent. The sequence of events in the cases of primary carditis is illustrated in Table XXIII.

Condition on Examination.

The present condition of 66 adolescents and young adults who had suffered from primary rheumatic carditis during childhood was determined as follows.

Heart Normal. This group, numbering 12 cases, is of special interest in that all the patients examined were found to have perfectly normal hearts and to be enjoying excellent health, despite the fact that when in hospital during childhood they were diagnosed as severe primary rheumatic carditis. The early records of these patients do not allow of any dubiety regarding the critical state of the heart, since they all exhibited well marked signs and symptoms indicative of considerable cardiac decompensation necessitating many weeks of complete rest in bed. Furthermore, it is interesting to note that on discharge from hospital a definite cardiac murmur was evident in every case. A point of great significance, however, is that none of these patients suffered from subsequent relapses of rheumatic infection and it would therefore appear that complete recovery may occur and cardiac integrity /

integrity be restored in children suffering from rheumatic carditis, provided that acute exacerbations do not supervene.

Organic Cardiac Lesion Present. Evidence of organic cardiac damage was found in 23 of the patients who reported for examination, representing 34.8 per cent. of the total. Fourteen of these patients were in comparatively good health and were able to be employed and to indulge in gentle outdoor sport without cardiac embarrassment. The remaining 9 patients were seriously crippled and were compelled to lead very restricted lives, being frequently confined to bed for long intervals. The various cardiac lesions found on examination were:-

Mitral Regurgitation . . 14 cases

Mitral Stenosis 8 cases

Aortic Regurgitation . . 1 case

Dead. It was ascertained that no fewer than 31 of the 66 cases of primary rheumatic carditis died during childhood. This represents a mortality rate of 47 per cent. Of the cases which died, 26 succumbed to the initial attack of carditis and 5 died during subsequent relapses. The probable explanation of the high death rate during the first attack is that many cases of primary rheumatic carditis remain undiagnosed for relatively long periods owing to the vague and insidious nature of the symptoms, with the result /

result that when eventually they reach hospital they are found to be in extremis.

The present state of the patients who suffered from primary rheumatic carditis during childhood is summarised in Table XXIV.

Recurrences. (PRIMARY CARDITIS)

<u>1st</u>	<u>2nd</u>	<u>3rd</u>	<u>4th</u>	<u>5th</u>	<u>No. of Cases.</u>
No Recurrences					50
Ca.	-	-	-	-	7
A.Ca.	-	-	-	-	1
Ca.	Ca.	-	-	-	3
Ca.	A.Ca.	-	-	-	1
A.Ca.	C.Ca.	-	-	-	1
C.Ca.	Ca.	-	-	-	1
C.Ca.	C.Ca.	C.Ca.	-	-	1
E.N.	E.N.Ca.	E.N.Ca.	E.N.Ca.	-	1
<u>Total</u>					<u>66</u>

KEY.

C.	-	Chorea.
Ca.	-	Carditis.
A.	-	Arthritis.
C.Ca.	-	Chorea with Carditis.
A.Ca.	-	Arthritis with Carditis.
E.N.	-	Erythema Nodosum.
E.N.Ca.-	-	Erythema Nodosum with Carditis.

TABLE XXIII. Showing the Sequence of Recurrences
in the Cases of Primary Carditis.

PRIMARY CARDITIS.

	<u>Cases.</u>	<u>Percentage.</u>
Normal	12	18.2
Mitral Regurgitation	14	21.2
Mitral Stenosis	8	12.1
Aortic Regurgitation	1	1.5
Dead	31	47.0
<u>Total</u>	<u>66</u>	

TABLE XXIV. Showing the Present Condition of the
Cases of Primary Carditis.

Summary of the Later Effects of Acute Juvenile
Rheumatism (All Manifestations).

In the foregoing pages the later effects of the three manifestations of acute rheumatism in childhood have been presented separately, and it will perhaps be useful at this stage to summarise the situation by a brief statement regarding the present condition of the total cases in the series.

The number of patients about whom complete information is available at the present time is 325 and, as previously stated, all of these who still survive have reached adolescence or early adult life, Heart Normal. In 144 cases, or 44.3 per cent., examination revealed the heart to be normal in every respect and the general health to be correspondingly satisfactory. The individuals included in this group were capable of full employment and were able to partake of strenuous sports without undue fatigue. Perhaps the most interesting cases which were eligible for inclusion under this heading were those in which active carditis had been present when they were in the wards of the hospital. It was a most gratifying experience to discover that these were now perfectly normal individuals who had in every way recovered their zest for working hard and playing hard./

Organic Cardiac Lesion Present. Under this heading 95 patients were classified, representing 29.2 per cent. All of these exhibited unquestionable signs of permanent cardiac damage, but subjectively there was a wide variation in the individuals examined, a considerable proportion being entirely symptom-free and living normal lives. This absence of subjective symptoms in many instances was very striking more especially when it was evidenced in association with gross cardiac signs. There can be little doubt, however, that the expectation of life of the patients in this group has been seriously curtailed and only a limited few will survive until the fourth decade.

Dead. Careful enquiry elicited the information that 86 cases in this series died before reaching the age of puberty. Of these, 81 are known to have died as the result of rheumatic carditis and 5 succumbed to other conditions unassociated with acute rheumatism. The actual "corrected" mortality rate for the whole series is 25 per cent. This figure clearly indicates that acute rheumatism, in addition to the serious crippling effects which it exerts on young children through its affinity for the heart, entails a grave menace to life, and when it is remembered that the revelations of this follow-up represent only the penultimate act in the drama, the tragedy of the final tableau /

tableau can scarcely be envisaged with complacency.

For ease of reference the above data regarding the total cases are presented in Table XXV.

TOTAL CASES.

	<u>Cases.</u>	<u>Percentage.</u>
Normal	144	44.3
Mitral Regurgitation	73	22.5
Mitral Stenosis	18	5.5
Aortic Regurgitation	4	1.2
Dead	86	26.5*
<u>Total</u>	<u>325</u>	

*Corrected Mortality - 25 per cent.

TABLE XXV. Showing the Present Condition of the
Total Cases in the Series.

SUMMARY of PART II.

The later effects of acute rheumatism in childhood have been investigated in a large series of cases, those selected for examination having reached adolescence or early adult life. The procedure of individual examination and interrogation of each patient was adopted in preference to the questionnaire method.

The three primary manifestations of acute rheumatism - chorea, arthritis and carditis - are discussed separately, and in each instance the recurrence rate, the sequence of recurrences, the present condition of the survivors and the death rate are recorded.

It transpired that 58.2 per cent. of the total cases suffered from recurrence of acute rheumatism. The highest recurrence rate occurred in the cases of primary arthritis where 71 per cent. are known to have suffered from subsequent acute relapses.

It is revealed that a child may recover completely from rheumatic carditis before adolescence is reached provided that recurrence of infection does not supervene in the interval.

Cardiac complications were discovered in 22.4 per cent. of the chorea cases and in 34 per cent. of the arthritis cases, the damage sustained by the heart /

heart being of a permanent nature. Of the patients who suffered from primary carditis, 47 per cent. failed to survive to adolescence, and a further 35 per cent. were found to be suffering from advanced cardiac damage.

Of the individual cardiac lesions detected, mitral regurgitation was the commonest. Definite signs of mitral stenosis were elicited in 5.5 per cent. of the total cases when they reported for examination.

The mortality rate for the whole series was 25 per cent. and a further 29 per cent. were found to have sustained permanent cardiac damage at the time when they reported for examination.

DISCUSSION.

It is probably no exaggeration to state that acute rheumatism in children presents one of the most baffling problems in medicine today. A great amount of work and careful investigation have been and are being done in an endeavour to find a solution of the problem, but, as yet, it has not been found possible to fit the pieces together in order to complete the picture.

Although the disease is far from being a product of modern conditions it is somewhat disquieting to find that the incidence is showing a definite tendency to rise in the area served by the Royal Edinburgh Hospital for Sick Children, namely, the South East of Scotland. This increasing incidence is not evident in all parts of the country, although reports issued from various large centres rarely record any appreciable decline in the disease. It is thus a matter of urgency that all available resources should be mobilised to combat this constant menace to the lives and health of children.

The modern trend in the investigation of acute rheumatism, and of other medical problems, is to concentrate /

concentrate more and more on laboratory methods of research and, at the present time, bacteriology is the science on which hopes are centred for the solution of the rheumatic problem. Such scientific study must, of course, continue and should be encouraged by every possible means, but there is perhaps a tendency on the part of clinicians and public health officials to overlook or neglect other aspects of the subject and to develop an attitude of despondency inspired by the belief that nothing useful can be done until the laboratory worker is able to furnish precise information concerning the etiology of the disease. It is surely important, therefore, at the present time to discover whether such an attitude as this is fully justified in the light of the knowledge which we already possess.

With juvenile rheumatism as with other ills which beset the human race, there is often an unfortunate tendency to defer any immediate action and salve the conscience by giving voice to the well-worn, but meaningless phrase - "something should be done". Such words are easily said, but to put precept into practice in this particular instance must constitute one of the most difficult problems of preventive medicine at the present time. The chief obstacle in the path of progress would appear to be our very incomplete understanding of the fundamental /

fundamental causative factors of the disease, and the defeatist who claims to believe that the present situation is incapable of betterment magnifies this obstacle until it assumes such proportions that a policy of indefinite delay seems entirely reasonable. But the important fact is apt to be overlooked that, although the etiology of juvenile rheumatism is imperfectly understood, we do possess a great deal of clinical knowledge about the disease and until full use is being made of such knowledge there can be no possible justification for an attitude of complacency. At the present time the problem of juvenile rheumatism and carditis is capable of approach from two angles, namely, alleviation and prevention, and these two aspects will be considered in turn.

Before discussing details of possible methods whereby practical help can be given to the rheumatic child it may be instructive to consider the treatment meted out to the tuberculous patient at the present time as compared with the benefits offered by the community to the child with rheumatic carditis. When a patient is found to be suffering from active pulmonary tuberculosis he is forthwith transferred to a specially equipped institution, the sanatorium, where he may remain for a period of months or, perhaps, years undergoing treatment under ideal conditions, and he is not permitted to leave the sanatorium /

sanatorium until such time as the disease is cured or completely quiescent. Furthermore, there is no time limit to the length of the patient's stay in the sanatorium, this being governed entirely by the progress made. After discharge, contact is maintained and the patient is usually subjected to frequent examinations by a specially trained medical officer and is instructed to report immediately at a special dispensary if he suspects any recurrence of symptoms. If any relapse has occurred the patient is promptly referred back to the sanatorium where he is detained for such time as is considered necessary by the physician in charge. The reason for this careful segregation and supervision of tuberculous subjects is, of course, twofold - benefit to the patient concerned and prevention of dissemination of the disease.

When a child develops rheumatic carditis he is usually admitted to a busy medical ward in a children's hospital where he may well remain for a period of several months. During a large part of his stay in hospital he is likely to be convalescent, yet he is occupying a bed in an institution which is intended primarily for cases of acute illness and which, more often than not, is sorely pressed for accommodation. Moreover, throughout his lengthy convalescence the unfortunate child is entirely deprived /

deprived of the mental stimulus of education, his sole interest being the somewhat morbid and frightening spectacle of the varied sufferings of other children in the ward. When at length he is discharged from hospital, perchance with a permanently damaged heart, he receives instructions to report back to the out-patient department or to an out-patient rheumatic clinic, which he may or may not do. If he fails to report then he is usually forgotten by the hospital until he returns for readmission on account of an acute relapse. If he obeys instructions and reports as an out-patient he is frequently given advice regarding such matters as rest, diet and avoidance of damp which is quite incapable of being acted upon in the type of surroundings from which he comes. Ultimately the unfortunate child may become a pupil at a "special school" where in the company of other children suffering from various types of disability he endeavours to acquire some elementary knowledge. Each morning he has to travel to school and each afternoon he has to return to his unsavoury home conditions, and the all too frequent outcome of this régime is an acute relapse resulting either in an untimely death or in further damage to an already overburdened heart muscle. This picture is no figment of the imagination but the result of considerable /

considerable personal experience.

To summarise the situation somewhat cynically, it may be said that the patient with phthisis is a danger to the community as well as to himself and, therefore, every concern is shown to him; the child with rheumatic carditis is a danger only to himself and so the community is largely unconcerned. In fact, it is probably no exaggeration to state that if acute rheumatism were proved to be highly infectious the lot of the rheumatic child would, as a result, be greatly improved.

It is sometimes argued that the expenditure of money on special institutions for cardiac invalids is unjustified since the disease cannot be cured by pleasant surroundings, and funds would be better directed towards an attempt to deal effectively with the early case. This attitude, which is not uncommon, would appear to be both shortsighted and callous. Furthermore, those who affect it are frequently found to be doing singularly little to help either the early case or the later case and the argument is inclined to be used as a convenient cloak for inertia. It is the hope of all who are seriously concerned with the ravages of juvenile rheumatism that the time will come when special institutions for cardiac children will no longer be required, but that time is not yet, and so long as children /

children continue to be crippled with heart disease, so long will it remain the duty of the community to provide for their needs. It is necessary, therefore, to consider carefully the question of the alleviation of suffering in young children afflicted with cardiac disability, and it may be of interest at this juncture to comment briefly on certain of the more ambitious attempts which are being made at the present time to deal with this aspect of the problem.

One of the most praiseworthy and enlightened schemes to aid the rheumatic child is fostered by the city of Birmingham. In 1921 the Local Education Authority opened the Baskerville Residential School at Harborne, on the south side of the city. This institution is reserved for children suffering from sub-acute and chronic rheumatism and contains 90 beds. The machinery whereby cases are selected for admission to the school constitutes an all too rare example of satisfactory co-operation between the public health service, the private practitioner and the voluntary hospital. Cases may be referred by the school medical service or by the private practitioner to the children's hospital, where they are examined by one of the hospital physicians who is also on the staff of the Baskerville School and who has the authority to select all cases for admission to the school. Such an arrangement is excellent since it /

it ensures that the final selection of suitable cases is made by an acknowledged expert in children's diseases and it necessitates but one clearing-station to which all cases are referred, namely, the children's hospital. Furthermore, the physician who selects the cases has the facilities for supervising their management whilst they are resident at the school, and is enabled to maintain close contact with the children after they have been discharged.

Whilst resident at a school such as Baskerville the rheumatic child receives daily education from a trained teacher and graduated exercises play an important part in the régime. The daily routine at this admirable institution is as follows:-

8 a.m.	Breakfast
8.30 a.m.	Play, rest, household duties.
9.20 a.m.	"Nerve Rest".
9.30 a.m.	Prayers, singing.
10 - 12 a.m.	Morning School.
12.5 p.m.	"Nerve Rest".
12.15 p.m.	Dinner.
1 - 2.15 p.m.	Resting, playing, walks in
2.30 - 4.30 p.m.	Afternoon School. garden.
4.35 p.m.	"Nerve Rest."
4.45 p.m.	Tea.
5.15 - 8 p.m.	Recreation and Bed. Time of
		going to bed depends on age
		or stage of convalescence.

During the intervals of "nerve rest" all children sit or lie silent with their eyes closed for a period of ten minutes, and are taught to relax all their muscles.

The above routine applies to the majority of children /

children in the school who, of course, have not been sent there until signs of active rheumatism have largely subsided. If, however, any child should show signs of exacerbation of symptoms, he is at once confined to bed until such time as the physician in charge of the school deems it advisable for him to resume the normal daily routine. The average duration of stay in the Baskerville Residential School is eight months, and after discharge every effort is made to maintain contact with the children and to guide and supervise their home lives.

Similar residential schools or convalescent homes exist at West Wickham, Broadstairs, Willesden and Hartfield in England and at various centres in America, the régime being conducted on similar lines in these institutions to that described for the Baskerville School at Birmingham.

Information regarding the results of prolonged convalescent treatment of rheumatic children is somewhat scanty and it is probable that some time will have to elapse before authoritative figures can be quoted. Poynton and Schlesinger (1937) who have experience of the home at West Wickham state: "sufficient time has not yet elapsed for any reliable statistics to have appeared on the results of prolonged convalescent treatment, but we are convinced of the beneficial effects. With patience most active /

active cases of rheumatism can be rendered quiescent. Not infrequently early cardiac lesions have disappeared entirely. . . . On the whole there seems to be general agreement that the majority of rheumatic children treated in this way improve and the incidence of relapse is low".

Since this work is concerned with rheumatic children in the Edinburgh area it is perhaps permissible at this stage to introduce a parochial note and to refer to the state of affairs which exists at the present time in the capital city of Scotland. It is unfortunately true that any such reference must, of necessity, be brief, since the almost negligible contribution made by the Edinburgh Public Health Authorities towards the mitigation of the rheumatic problem can be dismissed in a few lines.

In 1930 the Public Health Department instituted a children's rheumatic clinic for ambulant cases which meets on one morning each week at the Children's Hospital, and this sole gesture represents the sum total contributed by the City of Edinburgh towards a solution of the rheumatic problem. Despite eloquent appeals by Ritchie (1935) and others, the authorities remain unmoved and apparently indifferent, and the unfortunate Edinburgh child afflicted with rheumatic heart disease continues to drift /

drift in and out of hospital, his progress towards an untimely death hastened by official negligence. The accommodation available for children crippled with chronic heart disease at the Liberton Branch of the Longmore Hospital is both inadequate and unsuitable. The number of beds is extremely limited and the practice of segregating rheumatic children in company with children suffering from other chronic or incurable disease is almost universally condemned by all authorities on juvenile rheumatism.

A well-equipped convalescent home for children with rheumatic heart disease should be as much an integral part of the health services of all large urban communities as is a sanatorium for tuberculous cases. The type of convalescent home visualised would be situated in pleasant dry surroundings within easy access of the large hospitals, and the children would be enabled to live there in comparative comfort and happiness while enjoying the inestimable benefits of carefully supervised education and recreations. Such a scheme as this is surely amply justified on purely humanitarian grounds, and yet there is good reason to believe that it would also yield positive results. When following up the cases in this series, adolescents and young adults presented themselves for examination who were found to have perfectly normal hearts and yet when their hospital records /

records were scrutinised it was revealed that they had suffered from undoubted rheumatic carditis during childhood. I would like to state with emphasis that experience with large numbers of rheumatic children has convinced me that even severe cardiac damage can recover and recover completely and I believe that the number of such cases is capable of being greatly increased by the provision of prolonged convalescent treatment under ideal conditions. Owing to pressure on hospital accommodation there is an inevitable tendency for children with rheumatic heart disease to be sent home too soon, at a time when the disease appears to be quiescent and yet the general resistance may still be low. If instead of casting these children back into the very environment where they acquired the disease they were sent to the peaceful and hygienic atmosphere of a convalescent home, after spending some months in such an environment they must surely emerge far better equipped to withstand infection and to resist the ever-present menace of acute relapse.

The out-patient rheumatic clinic or cardiac clinic forms an important link in any rheumatic scheme and should be conducted within the precincts of the children's hospital, but alone such a clinic can fulfil no really useful purpose. At best it is merely a sorting-house to which doubtful cases may be /

be referred for diagnosis and established cases for transference to a suitable institution. The futility of the periodic bottle of tonic for the ambulant rheumatic child attending an out-patient clinic must be obvious to all who have studied the disease. Acute juvenile rheumatism cannot be treated in the out-patient department however elaborate the equipment or however skilful the personnel in charge, and, furthermore, the ward waiting-list has no place in the scheme of things. The rheumatic case which, on presenting itself at the out-patient clinic, shows any signs of active disease whatsoever, should be treated as an emergency and admitted to the hospital ward with as little delay as is permitted in a case of acute pneumonia. When at length all signs of activity appear to have subsided the child is transferred from the hospital to the convalescent school where he will remain for a period of several months and where every effort is directed towards building up the general health and resistance. In the fullness of time the child is sent home but close contact must be maintained and regular attendance at the rheumatic out-patient clinic encouraged. At the first indication of any recrudescence of the disease the cycle will have to be repeated. This régime would not necessarily prevent rheumatic heart disease but it might serve to check its advance and it would be a sure safeguard against /

against the unhappy and squalid invalidism and neglect which are all too frequently the lot of the rheumatic child today.

Details of administration of a scheme such as is outlined above would be easily and quickly settled if it were blessed with the whole-hearted and mutual support of the public health authorities, the voluntary and municipal hospitals and the family doctor. In this respect the situation existing in Birmingham is probably approaching the ideal.

It is perhaps not too much to hope for that, before very long, all large communities throughout Britain, including Edinburgh, will awaken to the realisation that the needs of the young child who, through no fault of his own, is struck down with rheumatic heart disease, are just as pressing as the claims of individuals afflicted with tuberculosis, syphilis or cancer.

In the foregoing pages suggestions have been offered which have as their aim the relief of distress and the delimiting of cardiac damage in the established case of juvenile rheumatism. When attention is given to the possibilities of effectively preventing rheumatic heart disease in children, this vast problem is found to be beset by the real and numerous /

numerous difficulties which encompass preventive medicine today, and it is their apparent magnitude which probably accounts for the fact that comparatively little is being done in an attempt to overcome them.

The main obstacles in the path of prevention would appear to be the early recognition of what has been termed the pre-rheumatic child, or of the child who is in danger of developing carditis, and the most satisfactory method of dealing with him when recognised, and in this connection it may be profitable to review certain of the points arising from this clinical study of the subject which, if borne in mind may assist in the anticipation of acute rheumatic infection in children and in the detection of potential cardiac disease.

Age Incidence. When a search is being made to evolve a plan for preventing or anticipating rheumatic heart disease in children it is advisable to consider the age incidence of the disease since the risk of infection appears to be much greater at some ages than at others. The child under five years is relatively safe, whereas the ages of seven and eight years represent the peak of the danger period necessitating the strictest supervision at this time. In girls a careful watch should be kept during the years immediately preceding puberty, but in boys this age /

age period is relatively safer and some relaxation of supervision is justified. It should be remembered, however, that boys are more prone than girls to develop primary carditis and for this reason the early recognition and treatment of apparently trivial signs of infection in boys is of prime importance.

Seasonal Incidence. Although acute juvenile rheumatism can occur at any season of the year it is, perhaps, important to bear in mind the fact that it tends to occur with greater frequency during the cold months, more especially in the first quarter of the year. This feature of the disease was clearly borne out in the present investigation, the incidence reaching its highest level in the month of January and its lowest in the month of August. Thus it would seem that the watch on the suspect child will require to be most concentrated during the cold winter months, the season when, apparently, the child is most vulnerable.

Family History of Acute Rheumatism. Information obtained in the course of this investigation affords strong evidence in support of the view that acute rheumatism in children is frequently a familial disease. If this is the case, then surely more might be made of this knowledge than is being done at the present time. The child of tuberculous parents or /

or grandparents is regarded with suspicion whenever the taint in his ancestry is discovered, and is rightly submitted to periodic examination in order to ensure detection of infection in its earliest stages. In the case of the rheumatic family no such precautions are taken, and the children of rheumatic parents are usually entirely ignored alike by the public health services and the hospitals. It may be argued that this apparent negligence is due to the fact that acute rheumatism is not a notifiable disease and that adequate supervision is thus impracticable; but it must be remembered that the vast majority of acute rheumatic cases, both children and adults, pass through the wards of the hospitals in the large cities and towns and through this source it should be possible to maintain contact not only with the patients themselves, but with other members of the patients' families who have, so far, escaped infection but who are, nevertheless, in danger of contracting the disease. In this respect a very strict watch should be kept on the child whose mother has suffered from acute rheumatism. Thus, if the familial tendency of acute rheumatism be borne in mind it may play an important part in the recognition of the early case.

Muscular Pains and Sore Throats. The significance of intermittent muscular pains and repeated sore throats /

throats in children cannot be doubted and yet these valuable warning signs are frequently ignored. This is probably due to the fact that these symptoms not infrequently occur in children who never develop acute rheumatism although in this investigation the proportion of these was found to be comparatively low. Since it is probable that recurring muscular pains in children are, in fact, evidence of rheumatic infection, it is surely difficult to exaggerate their importance and, if they occur in a child belonging to a rheumatic family, the failure to keep such a child under constant supervision is little short of culpable negligence. There is an unfortunate tendency when dealing with acute rheumatism, more than with other serious scourges, to wait until disaster is an accomplished fact before any active measures are taken. The child which suffers from frequent growing pains and tonsillitis walks constantly in the shadow of disaster, and if its heritage be bad the outlook may indeed be grave.

The Tendency to Relapse. It is well recognised that acute rheumatism is a disease which is very prone to recur several times in the same individual and that it differs from other acute infections in that one attack does not confer any immunity against subsequent attacks, but rather has the reverse effect, apparently rendering the child more susceptible /

susceptible to the disease. This liability to recurrence constitutes one of the most serious difficulties in the management of the rheumatic child, since it is quite impossible to foretell in any given case whether the first rheumatic attack will be the last or whether it is merely the forerunner of several similar or more serious illnesses. As has been noted in previous sections the recurrence rate in this series of cases was high although it showed considerable variation according to the nature of the initial acute manifestation, being highest in cases of primary arthritis.

The liability of the rheumatic child to suffer from acute relapses is a factor of great importance, but here again the attitude with which it is regarded is apt to be one of resignation. There is no reason to suppose that relapses are inevitable in acute rheumatism and, if not inevitable, then they are preventible. Not infrequently the heart is left unscathed after the first attack of acute rheumatism, and it is surely of vital importance that all possible measures be adopted to prevent a recurrence of the symptoms in the course of which irreparable damage to the heart may result. At the present time many children admitted to hospital with acute rheumatism are, in course of time, discharged as cured and are then completely lost sight of until they /

they return for re-admission on account of an acute relapse. This deplorable state of affairs was only too evident in the course of the follow-up of the children in this series, many of whom had been almost entirely ignored alike by hospital and public health service for several years.

If a serious attempt is to be made to prevent cardiac disease in children or to lower its incidence, then it is imperative that the knowledge of the strong tendency of acute rheumatism to relapse must rank as supremely important, since the prevention of relapses would frequently be tantamount to the prevention of heart disease. Further reference will be made to this important matter in later pages.

The observations presented in the foregoing paragraphs may be summarised in the following recommendations.

The frequent and thorough supervision of children who are in danger of developing rheumatic heart disease is an urgent need at the present time, but in order that this may be accomplished it is necessary to define what is meant by the child who is in danger.

Careful study of the subject would seem to indicate that a child of school age who is subject to muscular pains and repeated sore throats should always /

always be strongly suspect and if, in addition, there is a family history of acute rheumatism, the danger is thereby considerably increased. Bearing in mind the familial tendency of the disease, the children of rheumatic parents should be kept under strict supervision and examined at frequent intervals whether or not they exhibit symptoms of rheumatic infection.

The child who has suffered from one attack of acute rheumatism should never be permitted to drift beyond medical control, since the risk of recurrence is very great. Not infrequently serious cardiac damage is avoided at the first attack, but there is grave danger of this developing in the course of subsequent relapses.

The watch kept on the suspect child must never be relaxed, but during the cold winter months the vigil will require to be more constant owing to the greater susceptibility to rheumatic infection at this season of the year.

If it be possible in the course of careful examination and enquiry to discriminate between the child who is in danger of acquiring rheumatic carditis and the child who is not, then the vital question /

question at once arises as to whether any effective action can be taken to avert possible disaster. There are many who believe that with our present incomplete knowledge, juvenile carditis cannot be prevented, but it is only by challenging such a belief that possible ways and means will emerge to refute it. At the outset it has to be admitted that the path of prevention is strewn with innumerable obstacles but, great and formidable though these be, they yet may not be entirely insurmountable and the various approaches should be examined with determination and purpose.

If the many excellent schemes which are in operation both in this country and abroad to aid the rheumatic child be carefully examined and considered, it will be found that, although varying in detail, such enterprises have one common denominator in that they are all primarily concerned with the established case of rheumatic carditis. They are preventive in that they aim at checking the advance of the disease, thereby preventing renewed and more serious damage to the heart muscle, but they are mainly palliative and, as such, they cannot be the means of seriously reducing the incidence of heart disease in young people, although they may fulfil the very worthy purpose of prolonging life. There is a further limit /

limit to the usefulness of the schemes which are in existence at present, namely, the time factor, since even at the more ambitious of the special institutions for rheumatic children the length of stay has to be reckoned only in months owing to constant pressure on the accommodation. This entails sending the children home to an unsuitable environment when they are still of susceptible age and, although their resistance to infection has probably been raised by virtue of the convalescent treatment, the margin of safety is relatively small and the risk of relapse considerable.

When endeavouring to formulate a plan which aims at the prevention of rheumatic heart disease in children I believe that two fundamental conditions must be accepted to ensure any hope of success. The first of these conditions is that the children selected should have normal hearts and, secondly, the preventive measures adopted should be uninterrupted throughout the whole period of childhood. So far as I can ascertain, no rheumatic scheme in vogue at the present time fulfils these two conditions.

Certain points which may prove helpful in the selection of the child who, as yet unscathed, is in imminent danger of developing carditis, have already been indicated, but there is one important group on which, in the first place, all efforts might be concentrated /

concentrated, since the individuals who compose it are, beyond all doubt, walking in constant peril. I refer to the young children who have suffered already from acute rheumatism and have evaded enduring heart damage. In the earlier part of this work the great liability to acute relapse is made evident and the danger to which an undamaged heart is exposed in the course of such relapse is emphasised. Children conforming to this description, therefore, should take pride of place in any preventive scheme, since they are capable of selection with the minimal of difficulty and without any doubt as to their suitability. As previously noted, warning signs such as muscular pains and sore throats, and a rheumatic inheritance may play an important rôle in the recognition of early rheumatic infection and the anticipation of rheumatic carditis but, at the present time, an effective policy of prevention embracing all such cases is impracticable, since the necessary facilities could not be obtained to deal adequately with the very large numbers included under these headings. It were better, then, if whole-hearted effort were directed towards the prevention of recurrence of rheumatic infection since therein lies hope of preserving the integrity of the heart.

The putting into practice of a plan to prevent recurrence /

recurrence of acute rheumatism in children would only involve expansion of existing schemes in those more enlightened centres which are already endeavouring to cater for the rheumatic child, whereas in cities, such as Edinburgh, which are at present without any policy at all, considerable expenditure both of energy and money would be necessary to inaugurate and perpetuate an effective plan.

The complete rheumatic scheme envisaged would consist of four interdependent units, each constituting an important link in the chain of treatment and prevention. These units are:- the Out-Patient Rheumatic Clinic; the Hospital; the Convalescent School; the Residential School. To illustrate the functions of each of these units it will simplify matters to trace the progress of a rheumatic child from the time of the onset of acute rheumatism until the school-leaving age.

A girl aged nine years develops chorea and is referred by her family doctor or by the school medical officer to the Out-Patient Rheumatic Clinic at the children's hospital which serves the area. At this clinic the diagnosis is confirmed by the physician in charge who is, of course, a member of the permanent hospital staff and, after full out-patient records have been made the child is at once admitted to one of the wards for active treatment. Here /

Here she will remain until all symptoms have completely subsided or until the time when she would normally be discharged from hospital according to present day custom. When this stage is reached and while still an in-patient the child is referred back to the Rheumatic Clinic for re-examination by the same physician who admitted her in the first place, thus enabling him to complete his records of the case for future reference. The physician in question now has absolute authority to decide on the next step and he has three alternatives from which to choose. If he considers that there are still signs of active disease remaining, he may refer the child back to the ward for a further period of hospital treatment; if signs of activity are absent but there is a cardiac lesion present then the child is forthwith transferred to the Convalescent School; if the child is found to be normal in every way, arrangements are then made to have her admitted to the Residential School. The first of these alternatives is self-explanatory and would very rarely arise, but the second and third must be considered in more detail.

The child has recovered from chorea but is found to have a cardiac lesion. Under existing circumstances, such a child would be sent home from hospital and instructed to report at an out-patient clinic /

clinic at regular intervals. Under the proposed new scheme the child would not be sent home from hospital but would be transferred direct to the Convalescent School. This institution would provide a quiet, convalescent régime combined with educational facilities and would be modelled on similar lines to the Baskerville School at Birmingham. The minimum duration of stay would be six months but in many cases this might have to be exceeded. The Convalescent School would be essentially a haven for cardiac cases and the child with a normal heart would not be eligible for admission there.

To return to the child in question who has recovered from chorea but has been left with a cardiac lesion. She has passed by way of the Rheumatic Clinic through the Hospital out to the Convalescent School where every effort is made to increase her resistance to infection and build up the general health. At the end of six months of convalescent treatment she is again carefully examined by the physician who examined her in the first place, this time in his capacity as visiting physician to the Convalescent School. If, at this stage, the cardiac lesion is found to have healed, as may easily be the case, then it can be assumed that recovery is complete, but if the cardiac lesion persists unchanged, then /

then the probability is that it indicates damage of a permanent character and complete recovery is unlikely. The effect of these two possibilities on the future progress of the child will now be considered.

The child has been at the Convalescent School for a period of six months and at the end of that time a definite cardiac lesion still persists. In these circumstances the child will be sent home, provided that the visiting physician is satisfied with the state of her general health. The reason for this is the improbability that this child's heart will ever be completely restored to normal and, since she has had her chance, she must now make room for her more needy brothers and sisters. Before the child is sent home the nature of her illness is patiently explained to the parents and they are instructed to bring the child to the Out-Patient Rheumatic Clinic for examination once every month. From time to time the home is visited by health visitors and advice given regarding such matters as rest and general hygiene and, at the first sign of any recrudescence of infection, the child is at once re-admitted to the Hospital by way of the Rheumatic Clinic and the cycle is repeated.

This scheme, although far from ideal, would, I believe, have the effect of rendering the rheumatic child /

child less liable to serious relapse and would thus preserve cardiac efficiency for a much longer period. Furthermore, it has the virtue of ensuring that the child is constantly under expert supervision throughout the most vulnerable years.

The child has been at the Convalescent School for six months and her heart has completely recovered.

In this alternative event arrangements are made for the child to be admitted to the Residential School. This institution would resemble the Convalescent School in that it would be under strict medical control, but there the resemblance would end, since the Residential School would be concerned primarily with the education of healthy children and not with the treatment of rheumatism. But the fundamental principle of this, the preventive unit in the scheme, would be the absence of a time-limit, since the child would reside there until she had reached the school-leaving age and had put behind her the most crucial and dangerous years of her career. Thereafter, she would pass out of the school to her own home where she would have to take her chance in life, but I am convinced that the chance would then be infinitely better than if she had merely been discharged from hospital to her home environment after the original attack of acute rheumatism.

To return for a moment to the Out-Patient Rheumatic /

Rheumatic Clinic, the third alternative may be briefly considered.

The child has recovered from chorea in hospital and is found, on examination before discharge, to have a normal heart. In such a case the physician may refer the child direct to the Residential School omitting altogether the Convalescent School. His justification for so doing is that the child has emerged from her illness with her cardiac function unimpaired and is thus eligible for the Residential School. The effect of such a procedure would be to avoid undue pressure on the accommodation of the Convalescent School which, as previously explained, is intended primarily for children with cardiac disability and not for children with normal hearts.

And so we have followed the progress of the girl with chorea; from her home to the Out-Patient Rheumatic Clinic, thence to the wards of the Hospital, back to the Clinic and then out, either to the Convalescent School or the Residential School, according to the state of her heart. If she be fortunate, her goal is the Residential School where she will remain until her school days are ended; if she be less fortunate, then she will at least experience those health-restoring months in the happy atmosphere of the Convalescent School before returning to her home, and her parents will have the assurance that /

that, in the event of a relapse, their child will again be accepted into the peaceful and healthy surroundings which they themselves are unable to provide for her.

Only the skeleton structure of a plan has been presented, but details can readily be filled in once the framework is secure. Furthermore, the preventive aspect of the plan could be easily extended to include a wider range of rheumatic children than indicated, although at the beginning it would probably be advisable to concentrate on the prevention of acute recurrence in the child who has so far escaped cardiac injury.

The underlying principle and aim of a scheme such as has been outlined is to protect the rheumatic child from her home environment, and I feel certain that, unless steps are taken to ensure that such protection is offered, not for a few weeks or a few months, but throughout all the school years, the incidence of heart disease in young people will remain at its present high level and the toll of young lives will continue unabated. Juvenile rheumatism is so predominantly a disease of the child in poor circumstances that environmental influences must surely play a major part in its causation and, although we do not yet know which factor or factors are chiefly to blame, our knowledge is /

is sufficient to justify an attempt to reproduce for the poor child the conditions enjoyed by her more fortunate sisters belonging to the well-to-do classes. If I were to hazard an opinion as to the deepest gulf which divides the mode of life of rich and poor child, it would not be inadequate diet, or damp surroundings, or dirt; it would be disquietude. The child in poor circumstances lives in an atmosphere of everlasting noise which besets her by night as well as by day. Her hours of sleep are seriously curtailed and troubled by the close proximity of harassed voices or blaring wireless and she awakens unrefreshed and over-anxious to face the din of crowded streets and mass education. It is from such a peaceless existence that I would remove the rheumatic child and, until this be achieved, I can detect no gleam of hope for her.

One important advantage which would result from the provision of special institutions to accommodate rheumatic children would be the unique opportunity which would be afforded for intensive study of juvenile rheumatism in all stages of the disease. At the present time facilities for anything resembling team work in this direction are extremely meagre and the segregation of cases would be the best means of rectifying this unfortunate state of affairs.

The plan advocated, which aims at the limiting of /

of heart disease already present and, of even greater importance, the prevention of damaging relapses in rheumatic children, is no mere quixotic flight of imagination, but represents an attempt to define a practical policy which will have reasonable hope of achieving its ends. The expenditure of public money on "tinkering" with the problem of juvenile rheumatism is merely so much money wasted and it were far better either to abandon the fight altogether or to take new courage and launch out boldly without too carefully counting the costs.

In a national emergency the expenditure of vast sums of money is accomplished by a mere wave of the official wand. A disease which has been estimated to kill 12,000 and disable 60,000 persons each year in Great Britain must surely rank as such an emergency.

SUMMARY of DISCUSSION.

The opinion is expressed that there is evident today a lack of vision and enterprize regarding juvenile rheumatism in many quarters, and that such an attitude is not justified in the light of the knowledge which we possess about the disease. In this respect a contrast is drawn between the concern displayed towards the tuberculous subject and the relative unconcern shown to the child with rheumatic carditis.

A comprehensive scheme is constructed which has as its aims the alleviation of suffering in the established case of rheumatic carditis, the delimiting of cardiac disability, and the prevention of heart disease in a selected group of cases.

It is believed that the first two of these objectives could be attained by the provision of facilities for prolonged convalescent treatment under the most ideal conditions, and that such facilities should be an integral part of the health services throughout the country.

Having regard to the prevention of rheumatic heart disease in children, the view is expressed that, in the first place, effort should be directed towards /

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towards the prevention of recurrence of infection since, in many instances, permanent injury to the heart would, thereby, be avoided. It is believed that such an object would only be achieved by removing susceptible individuals from their home environment until they reach the school-leaving age.

The rheumatic scheme advocated consists of four units and the purpose of each of these is discussed. The scheme differs from such as are in operation at the present time in that it provides uninterrupted protection, throughout the most vulnerable years, for the rheumatic child whose heart is still intact.

REFERENCES.

- Atwater, R.M. (1927). Amer. J. Hyg., VII, 343.
- Baillou, Guillaume de, (1736). Opera Omnia Medica, Tom. IV, De Rheumatismo.
- Balfour, Wm. (1816). Observations on Rheumatism and Sprains. Edin., 1816.
- Barret, J.B. (1911). Tr. Roy. Acad. Med. Ireland, XXIX, 46.
- Benjamin, F.J. (1927). Lancet, I, 1175.
- Bertram, M. (1925). Brit. Med. J., I, 498.
- Bland, E.F. & Jones, T.D. (1935). J. Clin. Investig., XIV, 633.
- Boaz, E.P. & Schwartz, S.P. (1927). Amer. Heart J., II, 375.
- Bouillaud, J. (1836). Nouvelles Recherches sur le Rheumatism Articulaire, Paris, 1836.
- Bright, Richard (1831). Reports of Medical Cases, II, Lond. 1831, 468.
- Coates, U. & Thomas, R.E. (1925). Lancet, II, 326.
- Coburn, A.F. (1933). Amer. J. Dis. Child., XIV, 933.
- Coombs, C.F. (1927). Lancet, I, 579.
- Farnum, W.B. (1928). Amer. J. of Med. Sciences, CLXXVI, 474.
- Faulkner, J.M. & White, P.D. (1924). J.Amer.Med. Assoc., LXXXII, 425.
- Garrod, A.E. (1890). A Treatise on Rheumatism and Rheumatoid Arthritis, Lond. 1890.
- Hiller, R.I. & Graef, I., (1928). Amer. Heart J., III, 271.
- Horder, Lord (1926). Brit. Med. J., April 5th, Lumleian Lecture.

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REFERENCES (Cont'd.)

- Hunt, G.J., & Osman, A.A. (1923). Guy's Hosp. Rep., LXXIII, 383.
- Ingerman, E. & Wilson, M.G. (1924). J. Amer. Med. Assoc., LXXXII, 759.
- Lambert, A. (1920). J. Amer. Med. Assoc., LXXIV, 993.
- Mackie, T.T. (1926). Amer. J. Med. Sciences, CLXXII, 199.
- McSweeney, C.J. (1931). Arch. of Dis. in Child., VI, 367.
- Maddox, K. (1937). Med. J. of Austral., March 13th, 394.
- Medical Research Council, Spec. Rep. (1927). No. 114.
- Miller, R. (1926). Brit. Med. J. (Suppt.), II, 16.
(1927). The Medical Annual, p. 429.
- Ministry of Health Reports on Public Health and Medical Subjects (1927). No. 44.
- Naish, A.E. (1928). Lancet, II, 10.
- Pitcairn, David (1788). Quoted in Baillie's Morbid Anatomy, 2nd Edit. Lond., 1797.
- Poynton, F.J. & Paine, A. (1900). Lancet, II, 861.
- Poynton, F.J. & Paine, A. (1913). Researches in Rheumatism, London.
- Poynton, F.J. & Schlesinger, B. (1937). Recent Advances in the Study of Rheumatism, 2nd Edit. Lond., p. 26, 88.
- Poynton, F.J. (1925). Brit. Med. J., II, 788.
- Riesman, D. (1921). J. Amer. Med. Assoc., LXXVI, 1377.
- Ritchie /

REFERENCES (Cont'd.)

- Ritchie, W.T. (1935). Edin. Med. J., XLII, 117.
(1936). Brit. Med. J., I, 679.
- Robey, W.H. & Freedman, L.M. (1927). Boston Med. & Surg. J., April 14th, 595.
- St. Lawrence, W. (1920). J. Amer. Med. Assoc., LXXV, 1035.
- St. Lawrence, W. (1922). J. Amer. Med. Assoc., LXXIX, 2051.
- Scudamore, Charles (1827). A Treatise on the Nature and Cure of Rheumatism, Lond., p. 122.
- Starling, H.J. (1923). Guy's Hosp. Rep., LXXIII, 388.
- Sydenham, Thomas (1685). Works of Dr. Thomas Sydenham, tr. J. Swan, 2nd Edit. Lond. 1749, p. 245.
- Sydenham, Thomas (1686). Works on Acute and Chronic Diseases, tr. G. Wallis, Vol. II. Lond. 1788, p. 430.
- Thomson, A.P. (1925). Brit. Med. J., II, 794.
- Wallace, H.L. & Smith, A.B. (1934). Lancet, II, 1391.
- Wallace, H.L. & Smith, A.B. (1936). Edin. Med. J., XLIII, 452.
- Watson, Sir Thos. (1848). Lectures on the Principles and Practice of Physic, 3rd Edit., Vol. II, London.
- Wilson, M.G., Lingg, C. and Croxford, G. (1928). Amer. Heart J., IV, 917.
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